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Physical and Psychosocial Wellness among Adolescents with Hemoglobinopathic Disorders

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By

Naba'a Hussein Eidan

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Supervised by:

Prof. Nuhad Mohammed Aldoori ,PhD

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بسم الله الرحمن الرحيم

"يَرْفَعِ اللَّهُ الَّذِينَ آمَنُوا مِنْكُمْ وَالَّذِينَ أُوتُوا الْعِلْمَ دَرَجَاتٍ ۚ
وَاللَّهُ بِمَا تَعْمَلُونَ خَبِيرٌ"

صدَّقَ ٱللّهُ ٱلعَليُّ ٱلعَظِيمْ
سورة المجادلة: الآية 11

Dedication

I dedicate this achievement to my father and my mother, their love and effort have accompanied me in this process, without hesitating at any moment of seeing my dreams come true, which are also their dreams.

To my siblings & friends, who have been my support in the difficulties.

To the candle of hope that lit the path of researchers, ***Dr. Hussein Al-Ibrahimi*** *(Mercy and forgiveness for his soul), we have missed your presence so much, but you are alive in our minds. We ask God to make you a companion to his righteous saints in the gardens of bliss.*

Naba'a Hussein 2022

SUPERVISOR CERTIFICATE

I certify that this thesis, entitled **(Physical and Psychosocial Wellness among Adolescents with Hemoglobinopathic Disorders)** submitted by **Naba'a Hussein Eidan** and prepared under my supervision and guidance at the Department of Pediatrics, Faculty of Nursing, University of Babylon as partial fulfillment of requirements for the Degree of Master Sciences in Nursing.

Signature

Supervisor
Ph.D., Pediatric Nursing
Prof. Dr. Nuhad Mohammed Aldoori
College of Nursing
University of Babylon
Date: / /2022

Signature

Asst. Prof. Dr. Wafaa Ahmed Ameen
Head of Department of Pediatric Nursing
College of Nursing
University of Babylon
Date: / /2022

COMMITTEE CERTIFICATION

We the members of a Thesis Discussion committee, certify that we have reviewed the thesis **(Physical and Psychosocial Wellness among Adolescents with Hemoglobinopathic Disorders)** carried out by **"Naba'a Hussein Eidan"** and examined the researcher in its contents, and what is related to it on / / 2022

We decide that the thesis is accepted as partial fulfillment for awarding the degree of Master in Nursing Sciences with a specialty in pediatric Nursing and the estimate of ( ).

Signature
Member
Prof. Dr. Khamees B. Obaid
Date: / / 2022

Signature
Member
Dr. Mohammed R. Abd Ali
Date: / / 2022

Signature
Chairperson
Prof. Dr. Salma K. Jehad
Date: / / 2022

Approved by the council of the college of nursing

Signature
Prof. Dr. Amean A. Yasir
Dean of the College of Nursing, University of Babylon
Date: / / 2022

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Abstract

Hemoglobinopathy is a group of genetic disorders transmitted from parents to their children, spreads all over the world, and is characterized by a decrease or absence of the normal synthesis of hemoglobin, as in thalassemia, or the abnormal structure of hemoglobin as in sickle cell anemia, and it has an impact on physical and Psychosocial wellness, which leads to increased fears about physical appearance, and interference in the process of gaining independence and healthy relationships with family, friends, and society.

This study aim to assess physical and psychosocial wellness among adolescents with hemoglobinopathic disorders. Identify association between the physical and psychosocial wellness with certain demographic data. As well as to investigate the correlation in physical and psychosocial wellness regarding adolescents with hemoglobinopathic disorders.

A cross-sectional descriptive study design was used during the period from 19th October 2021 to 8th June 2022. This study was conducted in Babylon Province on (150) patients aged 10-19 years with hemoglobinopathic disorders at Babil Teaching Hospital for Maternity and Children. Data were collected by using an electronic scale, a non-stretchable measuring tape, and a modified questionnaire for the purpose of study, and analyzed electronically by using SPSS 26.

The study shows the overall physical wellness for more than half of the sample was fair, psychological wellness for more than half of the sample was well and social wellness for more than half of the sample was well. Some variables were associated with physical wellness, such as the patient’s age, educational level of patients and parents, the degree of consanguinity between parents, and hemoglobin level. Whereas, psychosocial wellness is linked to some variables, such as the patient's educational level, mothers occupation, economic status of family, type of illness, and complications. As well as there is a significant correlation between physical and psychosocial wellness at a p-value < 0.05.

The study concludes that there is a significant impact of hemoglobinopathies disorders on the physical and psychosocial wellness of adolescent patients. It is recommended to activate the roles of pediatric nurses and family caregivers in promoting and encouraging children with hemoglobinopathy disorders to achieve improved performance in their daily living activities, psychological wellness, and social relationship, Particularly for children with little social support and high severity of sickness to increase their awareness by taking care of them in all aspects that affect and weaken their personalities.

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No.	Item	Meaning
1.	ABG	Arterial blood gases
2.	Ass.	Assessment
3.	BMI	Body mass index
4.	BMT	Bone marrow transplantation
6.	CO_2	Carbon dioxide
7.	DF	Degrees of freedom
8.	*et al.,*	And others
9.	F	Frequency
10.	g/dL	Gram per deciliter
11.	G6PD	Glucose-6-phosphate dehydrogenase deficiency
12.	H_0	Null Hypothesis
13.	H_1	Alternative Hypothesis
14.	Hb	Hemoglobin
15.	HbA	Adult hemoglobin
16.	HbA_2	Minor Adult Hemoglobin
17.	HBB	Hemoglobin subunit beta
18.	HbC	Hemoglobin (C)
19.	HbD	Hemoglobin (D)
20.	HbE	Hemoglobin (E)
21.	HbF	Fetal hemoglobin
22.	HbS	Hemoglobin (S)
23.	HCV	Hepatitis C virus
24.	HgbS	Hemoglobin (S)
25.	HgbSC	Heterozygous SCD patients (typically milder form)
26.	HgbSS	Homozygous SCD patients (most severe form)
27.	HLA	Human leukocyte antigens
28.	HRQoL	Health related quality of life

29.	HSCT	Hematopoietic stem cell transplantation
30.	IQ	Intelligence quotient
31.	M	Mean
32.	MENA	Middle East and North Africa
33.	MRI	Magnetic resonance imaging
34.	N	Number
35.	NBS	Newborn screening
36.	ng	Nanogram
37.	No.	Number
38.	O_2	Oxygen
39.	P_{50}	Partial pressure at which the hemoglobin molecule is half saturated with O_2
40.	PGD	Preimplantation genetic diagnosis
41.	PMS	Premarital screening
42.	PND	Prenatal diagnosis
43.	PO2	Partial pressure of oxygen
44.	r	Person correlations
45.	RBC	Red Blood Cell
46.	RCTs	Randomized controlled trials
47.	**SCA**	Sickle cell anemia
48.	SCD	Sickle cell disease
49.	SD	Standard deviation
50.	Sig	Significant
51.	SOB	Shortness of breathing
52	SPSS	Statistical Package for the Social Sciences
53	TDT	Transfusion-dependent thalassemia
54	TMT	Thalassemia major trait
55	US- FDA	United States- Food and Drug Administration
56	US$	American dollar
57	VOC	Vaso occlusive crisis
58	WHO	World Health Organization
59.	X^2	Chi-square
60.	α	Alpha

61.	β	Beta
62.	β +	Present of β-globin
63.	β0	The complete absence of production of β-globin
64.	β-TM	Beta Thalassemia Major
65.	Eγδβ	Epsilon-gamma-delta-beta thalassemia
66.	%	Percentage
67.	γ	Gamma
68.	δ	Delta
69.	<	Less than
70.	>	More than
71	≤	Equal or less than

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CHAPTER ONE

INTRODUCTION

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INTRODUCTION

1.1. Introduction

Haemoglobinopathies are the most frequent single-gene disorders worldwide, with populations from southern Europe, the Middle East, Africa, Asia, and the Pacific having a higher prevalence. They are defined by the absence, reduction, or abnormal synthesis of globin chains, and can be linked to broad range of phenotypic manifestations, including intra-uterine fetal death to the condition of asymptomatic carrier (Borbolla Foster *et al*., 2021).

Hemoglobinopathy as umbrella term which includes all hereditary hemoglobin disorders, this may be either cause qualitative defect such as sickle cell anemia or causing quantitative defect such as thalassemia (Abd *et al*.,2020).

The inherited nature of this disease beginning in childhood, changes in appearance, the likelihood of early death, and the necessity for ongoing therapy have a significant impact on the mental and psychological health of patients and their families. So, all patients with hemoglobinopathies or other chronic medical conditions are at risk for psychological problems (Sadeghloo *et al*. 2019, Zolaly *et al*., 2020).

The effect of disease on physical health leads to physical deformity, growth retardation, and delayed maturity. As well as, it impacts physical appearance, e.g., short stature, bone deformity, and also contributes to a negative self-image. Severe problems such as cardiac failure, arrhythmia, hepatic disease, endocrine problems, and infections are common among thalassemic patients (Behdani *et al*.,2015).

There is a well-established link between chronic diseases and psychological issues, cause hemoglobinopathic defects considered as chronic blood illness that requires time-consuming treatment, sufferers are more prone to experience psychological health problems (Thiyagarajan *etal*.,2019).

Sickle cell disease (SCD) as one of these defects; is reflected as a genetic blood illness that can have a negative impact on a person's physical and psychosocial well-being, including increased rates of depression and anxiety symptoms (Reinman,2019).

Children with SCD have a reduced physical, psychological, and social health-related quality of life, as well as restrictions in self-esteem, school and social participation, and overall psychological adjustment, all of which contribute to poor academic achievement (Abid *etal*., 2019). Negative emotions were visible, especially during painful crises (Foster & Ellis, 2018).

In sickle cell disease the falsified red blood cells can clog the microcirculation, leading to ischemia reperfusion damage, inflammatory cytokines, discomfort, and functional impairment, especially in the musculoskeletal system, which can lead to defensive inactive behavior even in young patients. It has previously been shown that childhood physical activity (PA) improves health in both the short and long term, and may lower risk factors for chronic diseases (Melo *et al*., 2018).

Other defected hemoglobin as thalassemia also has a significant negative effect on the patients' wellness. Particularly the impacted children are subjected to numerous stressful events throughout their lives, include frequent blood sampling for laboratory tests, multiple transfusions, recurrent subcutaneous injections, and oral iron chelator drug therapy, all of which make patients vulnerable to psychiatric burden such as anxiety and depression. Furthermore, limitations in social activities, fear, pain, and concerns about diagnostic procedures and transfusions every 2 _ 4 weeks, all of which cause stress, lead to sickness absence from school and poor educational performance. According to many research, psychological disorders are more common in people with significant thalassemia, with over 80% of them suffering from at least one mental disorder. According to the data, the most common problems were self-mutilation, anxiety, and serious depression (Pattanashetti *et al*., 2017; Batool *et al*.,2017).

Clinical manifestation of thalassemia major appears between the ages of 6 and 24 months. A β-Thalassemia major child's earliest symptoms include failure to thrive and a gradual paleness. feeding issues such as frequent diarrhea, irritability, fever, and abdominal distention occurs due to enlargement of liver and spleen. Long bone deformity and typical craniofacial changes such as skull bossing, mongoloid slant of the eye, depression of the nasal bridge, and enlargement of the maxillae leading to expose of upper teeth are among the skeletal deformities (Nigam *et al*., 2017; Cappellini *et al*., 2021).

Adolescent period is transition from childhood to adulthood. This is a dynamic phase with the following characteristics: rapid physical, physiological, psychological, and social development, growth, and maturity. Physical, mental, emotional, social, cultural, moral, professional, self-esteem, and identity-related transformations occur during adolescence; as this stage needs a healthy lifestyle based on support with a high level of nutrition and psychological and emotional stability (Özdemir *et al*., 2016).

A chronic disease affects all parts of a patient's life, not just the physical, but also the social and emotional, educational, and occupational aspects (Colombatti *etal*., 2021). Adolescent period is a critical stage of life so thalassemic adolescent undergoing for alteration in perception because of repeated blood transfusions, severe complications, school absenteeism and psychosocial burdens (Issa *et al*., 2018).

The emotional and cognitive needs of a child with hematologic disease are considerably different from those of an adolescent seeking independence and identity (Pranandita ,2021).

Patients with hematologic disorders suffer from biopsycho-social, and culture issues despite they are not initial causes, however, health care provider particularly nurses have noted that those patients are more likely to be stressed especially those with sickle cell anemia and thalassemia (Aldossari,2019).

Psychosocial difficulties that affect one's quality of life are becoming a more prominent focus of attention. Chronic disorders are well understood to have psychosocial consequences, which are especially important in children. Children with chronic medical ailments, such as thalassemia, are more likely to develop emotional and behavioral disorders (Behdani *et al*. 2015).

These patients require additional psychological support in order to reduce emotional stress, build competence, and promote therapeutic compliance in daily life (Pranandita, 2021).

Nurse's most important role is to support patient to take an active part in providing care and meet their requirement not only medically but also psychologically. Nurses are also play essential role in controlling a patient's quality of life by preventing additional complications and delivering therapy that minimizes interference with school or work commitments (Azize *et al*., 2015).

The impact of pain and symptoms on sickle and thalassemic patients and their families' daily lives, as well as society's attitudes toward disease and those affected, are the main causes of psychosocial concerns for patient and their families. Lack of social contacts, particularly from friendship and social activities, was a major source of dissatisfaction. A person's capacity to make friendships and social networks will have a significant impact on their ability to cope in society (Aldossari,2019).

Coping strategies with pain and the related complex treatment regimen e.g., recurrent hospitalization is frequently linked to comorbid psychic symptoms, impaired family functioning, chronic fatigue, and sleep disturbances in patients, all of which can be aggravated by socio - economic and environmental issues that might be difficult to manage. Given the etiology of pain in children with disease, evaluating morbidity needs a biopsychosocial integrated approach to assessing, defining, and managing pain (Hood *etal*.,2021).

1.2 Importance of Study

Hemoglobinopathies are the most common single - gene disease and one of the most serious health problems worldwide. About 2–7% of people have inherited hemoglobin diseases caused by quantitative or qualitative anomalies in the hemoglobin molecule (Karamperis *et al*.,2021).

The global statistic of hemoglobin diseases including around 7% of the global population. Hemoglobin disorders also affect 300,000 to 500,000 children annually, 70% of these children are born with sickle cell anemia, with the remaining 30% having thalassemia. The mortality rate of Sickle cell anemia is 50-80% for children every year, while thalassemia major kills 50,000–100,000 children each year (Ministry of Health – Kingdom of Saudi Arabia, 2022).

The incidence and prevalence of chronic illness is more common in both industrialized and developing countries and will constitute the major cause of death particularly among adolescents (Abdulsattar & Hattab, 2017).

SCD has been related to a high rate of morbidity and mortality, especially in children under the five years of age and teenagers. It can also have negative social and economic implications (Pandarakutty *et al*., 2020).

Every year, around 60,000 newborns are born with a significant β-thalassemia disorder, according to WHO reports. A deficiency in the β-globin gene on chromosome 11 causes β-thalassemia, which is a more serious clinical condition than α-thalassemia (Töret *et al.* ,2019).

In the United States the prevalence of β-thalassemia major is approximately 2,000 individuals. And the prevalence of SCD is 7–10% of African Americans (kliegman *et al*., 2020).

In Nepal, the prevalence of different hemoglobinopathic disorders is estimated to be 26.8% for thalassemia trait and 21.6 percent for SCD (Shrestha *et al*.,2020).

In India, the overall prevalence of thalassemia trait and sickle cell hemoglobinopathy is 3–17 percent and 1–44 percent, respectively (Gupta *et al*., 2018). SCD is the second most common hemoglobinopathy, after Thalassemia. The prevalence of hemoglobin S is 4.3% (Lakhani *et al.* ,2021). Whereas in Southern Iran, β -Thalassemia the most common prevalence mutation approximately 31% (Nezhad *et al*., 2018).

β-Thalassemia, SCD, and other hemoglobinopathies are regarded to be the most common autosomal recessive blood disorders worldwide, as well as in Middle Eastern countries. By 2050, the number of children born with SCD is anticipated to rise by 30% globally. According to available evidence, congenital and genetic illnesses are responsible for a significant share of mortality and handicap in this region, resulting in a poor quality of life for youngster. In Middle Eastern countries, the prevalence of hereditary blood disorders is entwined with social, cultural, and religious behaviors, making treatment more difficult. This places a significant burden on individuals, families, and health-care systems in these countries (Natarajan & Joseph, 2021).

In Saudi Arabia, the prevalence rate of β-thalassemia and SCD in examined populations showed a larger incidence of SCD than β-thalassemia major: 0.38 percent and 0.07 percent, respectively (Al Alwan *et al*., 2021).

In Syria, the prevalence of the β-thalassemia traits is estimated to be around 6.0 percent of the population, with an estimated 779,000 carriers and over 8000 transfusion-dependent patients (Yassouf *et al*., 2019).

Iraq has a monthly birth rate between (100-150) children effected with disease, which equates to around (2000) instances each year, and these results are disastrous when compared to neighboring countries (Alsudani *et al*., 2017).

In Iraq, between 6-10% of the population has hemoglobinopathy, of which thalassemia is a major component (Faraj *et al*., 2017). Furthermore, illustration in Najaf, the frequency of hemoglobinopathic disorders is 75.6 per 100,000 people. Major thalassemia (33.15%), intermediate thalassemia (31.91%), and sickle thalassemia (12.65%) are the three most common type of disease (Al-Hakeim *et al*., 2020). As well as the prevalence rate of thalassemia in Thi-Qar is more than other type of hemoglobinopathies 76.53% (Abd *et al*., 2020).

In the north of Iraq Hemoglobinopathies are a serious health issue among Iraqi Kurds. These illnesses represented as thalassemia is the most common, with carrier rates ranging from 3.7 to 6.9%. Alpha thalassemia is less common, with carrier rates ranging from 0.03 to 1.22 %, but the sickle cell gene has a wide range of carrier rates, ranging from 0.06 to 1.2 % (Al-Allawi *et al*., 2021).

According to a survey published by the Thalassemia International Federation (TIF) in 2008, just about 200,000 thalassemic patients are living over the world, and recorded as receiving a regularity treatment. The cost of patient management in Iraq, according to a 2016 evaluation by the Federal Board of Supreme Audit, is US$1428.00_3785.00 per month. In Iraq, epidemiologic information about disease burdens, complication, and other features are limited (Kadhim *et al*., 2017).

The management of SCD and TM has a significant cost impact on national health systems and local population quality of life, necessitating rapid public health action (Al-Riyami&Daar,2018).

There are several barriers to seeking care for patients, but one of the most powerful and frightening is stigma. People with hemoglobinopathy may face health stigma for a different cause such as race, socioeconomic level, illness conditions, delayed of puberty and growth, and having acute or chronic pain that requires to be managed with opiate. stigmatizing social encounters can cause negative consequences on psychosocial wellness such as social isolation, Internalized stigma, anxiety, depression, and suicide thoughts and attempts (Bulgin *et al*.,2018).

1.3. Problem Statement

Physical and Psychosocial Wellness among Adolescents with Hemoglobinopathic Disorders

This study is conducted on the physical and psychosocial wellness of hemoglobinopathic patients to identify the extension of diseases and effect on their daily living activities, school performance, behaviors, emotions and relationship with their friends, families and community.

1.4. Objectives of the Study

1. Assess physical and psychosocial wellness among adolescents with hemoglobinopathic disorders.

2. Identify association between the physical and psychosocial wellness with certain demographic data.

3. Investigate the correlation in physical and psychosocial wellness in regard to adolescents with hemoglobinopathic disorders.

1.5. Hypothesis

H_0: There is no significant correlation between physical and psychosocial wellness among adolescents with hemoglobinopathic disorders.

H_1: There is a significant correlation between physical and psychosocial wellness among adolescents with hemoglobinopathic disorders.

1.6. Definition of the Terms

1.6.1. Physical Wellness

a. Theoretical Definition

Physical wellness is defined as a consist of recognition the need for physical activities, healthy diet and sleeping as well as prevention illness and injury or managing chronic health problems (Northwestern university, 2022).

b. Operational Definition

physical wellness can be attributed to how hemoglobinopathies disorder effects on the patient's ability to perform activity of daily living and degree of satisfaction about physical appearance.

1.6.2. Psychosocial Wellness

a. Theoretical Definition

Psychosocial wellness is a multidimensional structure including psychological, sociological, and subjective elements that affect a person's total functionality in realizing their full potential as a member of the community (Kumar, 2020).

b. Operational definition

It Involving both social and psychological components. Psychosocial experiences can be attributed to how hemoglobinopathies disorder effects on the patient's psychological (emotions, feelings and thoughts) and social lives.

1.6.3. Adolescents

a. Theoretical Definition

Adolescence is a phase of development and progress between childhood and maturity. According to WHO considers anyone between the ages of 10 and 19 to be an adolescent (Csikszentmihalyi, 2021).

b. Operational definition

It is a transitional period between childhood and adulthood, this stage characterized by hormonal changes and intellectual, physical, social and psychological growth and development. Also, individuals achieving independence at this stage.

1.6.4 Hemoglobinopathic Disorders

a. Theoretical Definition

Hemoglobinopathies are a type of genetic blood disease that affects the structure, function, or generation of the hemoglobin molecule (AlDeen *et al*., 2021).

b. Operational Definition

Hemoglobinopathies include inherited groups of hematologic disease passed from parents to children. This disorders effect on the hemoglobin molecule either causing structural defect (deform of blood cell to abnormal shape) in state of sickle cell disease or causing functional defect (reduce or absence of synthesis of globin chains) in state of thalassemia. These disorders cause various degree of hemolytic anemia from mild to life threatening.

CHAPTER TWO

LITERTURE REVIEW

CHAPTER TWO

LITERTURE REVIEW

This chapter presents a logically and systematically organized summary of literature and studies relevant to the phenomenon investigated which are systematized in the following manner.

2. 1. Theoretical Concepts of Hmoglobinopathic Disorders

Hemoglobinopathies are diseases caused by genetic mutations that produce substantial structural alterations in the hemoglobin molecule. This leads to circulatory system obstruction due to changes in form, oxygen-carrying capacity, or the ability to cluster together. Thalassemia syndromes and structural hemoglobin variations are the two basic types of hemoglobinopathies. The most common kinds of thalassemia are alpha (α)- and beta (β)-thalassemia; the structural hemoglobin variations are HbS, HbE, and HbC. Each category contains numerous subtypes and mixed forms (Moussa& Chen, 2021).

Inherited hemoglobin disorders, basically SCA and beta-thalassemia are mostly prevalent monogenic diseases around the world. The prominence of this hematologic disturbance can't be overemphasized, approximately (5 to 7percent) of the world's population carries significant hemoglobin mutation. sickle cell disease is often depicted as the (first molecular disease) after the discovery of Linus Pauling in 1949 that the defect was caused by a malfunction of the hemoglobin molecules (McGann *et al*., 2017).

Thalassemia is a Greek word derivative from 2 words: Thalassa, which means sea, and Emia, which means blood, and is also known as Mediterranean anemia or Cooley's anemia in Persian. Thalassemia is a congenital hemolytic disorder that follows Mendel's rules of inheritance. In 1925, an American scientist named Dr. Cooley was the first to define it (Tari *et al*., 2018).

Hemoglobinopathic disorders were first reported from Iraq in the mid-1960s of the previous century (Al-Allawi *et al*., 2021).

2. 2. Epidemiology

Hemoglobinopathies are genetic blood disorders that are characterized by varying degrees of anemia and a wide range of clinical manifestation. They are most common in the Mediterranean region, Sub-Saharan Africa, the Mideast, Central India, and Southeast Asia, where an estimated 400,000 babies are born with significant hemoglobinopathies each year. Ninety percent of these births take place in de developed or development countries (Alzahrani *et al*.,2020).

Hemoglobinopathies and thalassemia continue to be a major cause of morbidity and mortality around the world. Although historically limited to endemic malaria regions, people with hemoglobin abnormalities are now found all over the world, and laboratories in previously in malaria free areas, such as North America, northern Europe, Japan, and Australia, are now being to diagnose hemoglobinopathies and thalassemia. Hemoglobinopathies are the most common genetic diseases in the world, especially in areas where malaria is widespread (Sabath, 2017).

α- and β-thalassemia, SCD, HbE disease, and HbC disease are all clinically relevant hemoglobinopathies. More than 330,000 births are predicted to be affected each year, majority with SCD (83%) and the thalassemia (17%)(Goonasekera *et al*., 2018).

SCD is one of most prevalent genetic illnesses in the world, affecting about 100,000 Americans, as well as people of other ethnicities like African, Hispanic, Italian, Greek, Iranian, and Turks, as well as those of Arab, Caribbean, and Asian Indian origin. The disease's prevalence varies depending on where you live. Sickle cell trait affects around 8% of African Americans, although it affects up to 40% of people in West Africa (Hockenberry *et al*., 2019).

In the United States, one out of every 400 Black infants have sickle-cell disease. In the United States, about 1 in 12 Blacks had sickle cell traits (a child carried the gene for the disease but does not have active symptoms) (pillitteri, 2018).

According to the WHO, around 20–25 million people globally have homozygous SCD, with 12–15 millions in Sub-Saharan African, 5–10 millions in India, and about 3 millions in other areas of the world. Annually, it is predicted that 312,000 people with sickle hemoglobin are born around the world, with the majority (236,000) in Sub-Saharan Africans, and the sickle cell gene having a gross prevalence of 6% in Oman. The annual increased in newborn cases is approximately 120–150 (Alkindi *et al*., 2021).

The higher frequency of consanguineous marriages, especially between the first relatives, is likely to be responsible for the higher incidence of SCD in Middle Eastern countries. Saudi Arabia (42–67%), Sudanese (44–63%), Qatari (54%), the United Arab Emirates (40–54%), Jordon (29–64%), Yemeni (40–45%), and Egyptian (21–33%) have all reported high percentages of consanguineous marriages. Eastern Saudi Arabia has the highest incidence of sickle cell disease and sickle cell trait in the Middle East (2.6% and 21%, sequentially) (Pandarakutty *et al*., 2020).

Sickle cell anemia, and TDT also recognized as major thalassemia, are compound hemoglobinopathies frequent in Omani, with approximately carrier incidence rates of 5.8% and 2.2 % sequencely. Sickle cell disease has a prevalence rate of 3.7 per 1,000 live births, and in a community-based study of 6,342 children, incidence rates of Sickle cell disease and β- thalassemia were found to be 0.2 % and 0.07 %, respectively, among children under the age of five (Al-Riyami & Daar, 2018).

Thalassemia is a hereditary disease resulting from mutations in the β-globin (chromosome 11) or α-globin (chromosome 16) genes, which causes a decrease of the affected globin chain and the buildup of the normal one, resulting in globin chain imbalance. As a result, inefficient erythropoiesis, erythroid membrane damage, and splenic blockage might ensue, accelerating hemolytic anemia or hematopoiesis problems. Thalassemia is a major health disease that affects people all over the world, particularly in the Mediterranean, Southeast Asia, and Southern China (Lee *et al*., 2019).

One of the most frequent autosomal recessive illnesses in the world is beta-thalassemia major. It's marked by a lack of hemoglobin production and a high rate of red blood cell destruction. It is present in over 60 countries, with a particularly high prevalence among youngsters in the Mediterranean, the Middle East, especially Iraq, and central and south Asia (Yousif &Yacoub, 2019).

Thalassemia is a public health issue in many parts of the Middle East and Northern Africa, Southeast Asia, Central Asian, and India. The prevalence of -thalassemia and carriers is higher in the MENA area, particularly in the Gulf

Cooperation Council (GCC) countries and the United Arab Emirates (UAE) (Shivappa *et al*., 2021).

Thalassemia is common within Southeast- Asia, but rare in Korea even so, the number of Southeast Asian immigrants to Korea has recently increased. Because of the growing number of Southeast Asian immigrants in Korea, the prevalence of thalassemia in young people is rising. Every year, over 56 thousand babies were born with serious β- or α- thalassemia, with more than 50% of them requiring constant transfusions. Hydrops fetalis, which is caused by α-thalassemia major, causes about 5,500 prenatal deaths each year. Thalassemia patients and carriers are now found all over the world as a result of population migration and change in recent years (Lee *et al*., 2019).

In Iran, where there are more than three million beta-thalassemia carrier genes and roughly 800 children born with thalassemia are added to this group each year, the prevalence rate of thalassemia is around 3–4% (Shahraki-Vahed *et al*.,2017).

Thalassemia is also most common in Arab countries with different recurrences where Iraq demonstrated difference in the prevalence of β-thalassemia in different areas from 3.7% to 4.6%. Recently, it was found that, severe β-thalassemia rates for 50,000 to 100,000 deaths yearly among children below the age of five in low and middle economic nations and in Iraq it was found that, there were more than 2,000 cases of thalassemia in the Kurdistan area and Kirkuk with approximately 30000 individuals are carries of β-thalassemia disorder (Tawfeeq, 2018).

Thalassemia is one of the common hereditary hematologic diseases in Iraq, with a predicted 15,000 registered patients with major and intermediate thalassemia, and an average dispersion of carriers of approximately 4% (Shareef & Obaid, 2017).

2.3. Structure, Function and Type of Hemoglobin

Hemoglobin is protein that is abundance in RBC and acts as a major O_2 transporter. Which delivers O_2 from the lungs to the body tissues and return O_2 to the lungs in the form of CO_2 (McGann *et al*., 2017).

During fetal life, hemoglobin in erythrocytes differs from that generated after birth that drops to 1% of adult levels by six months of age. Fetal hemoglobin is made up of 2α and 2γ polypeptide chains. This type of hemoglobin represents 40 percent to 70 percent of an infant's hemoglobin at birth (HbF). Adult hemoglobin (hemoglobin A), which is formed of 2α and 2β chains, increasingly replaces this throughout the first six months of life. As a result, diseases like sickle-cell anemia and thalassemia, which are β chain abnormalities, do not manifest clinically till HbF alteration has happened (at approximately 6 months of age). This can be detected prenatally by analysis of the hemoglobin or electrophoresis in fetal or newborn life since a little HbA is present during early intrauterine life (Pillitteri, 2018). Represented in the figure below

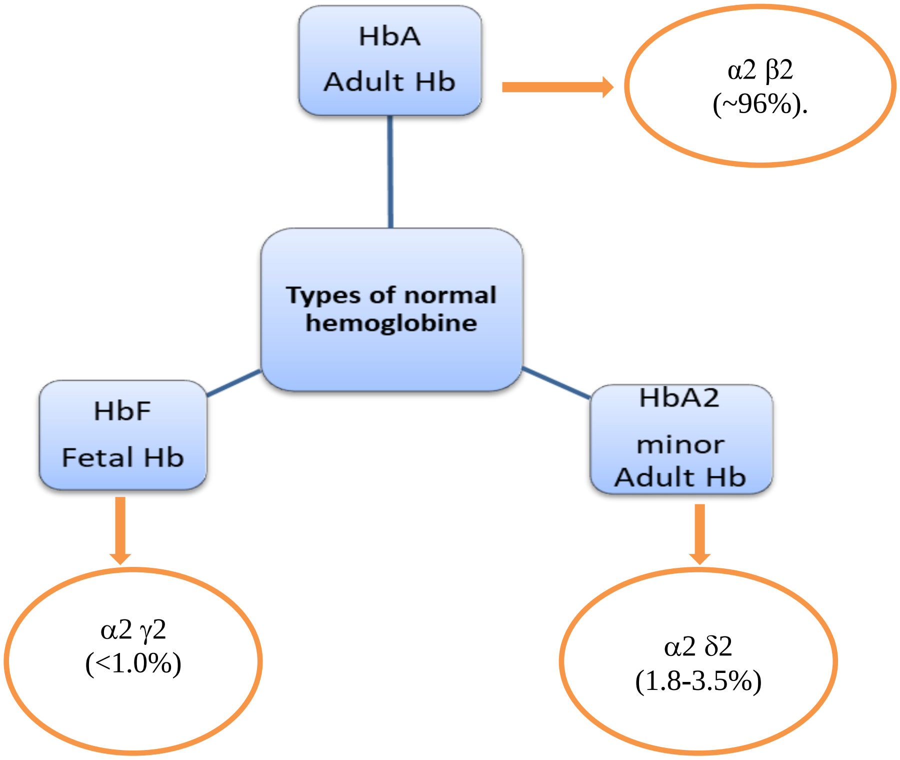

Figure (2-1) Types of normal hemoglobin after 6 months of age (Al-Allawi, 2017).

There are over 800 different types of hemoglobin which is the most prevalent and useful clinical classification of hemoglobinopathies. There are two Hb gene clusters, positioned at the ends of the shortened arms, for chromosomes 11 and 16, that are involved in Hb production. Their regulation is complicated, incorporating a location upstream regulatory area on each chromosome as well as an X-linked control site . The alpha (α) gene cluster on chromosome 16 contains three genes: zeta (ζ), alpha 1 (α1) and alpha 2 (α2). The beta (β) gene cluster on chromosome 11 contains five genes: epsilon (ε), gamma 1 (γ1), gamma 2 (γ2), delta (δ), and beta (β) (Kliegman, 2020).

2.4. Hemoglobinopathic Disorders

Hemoglobinopathy refers to a group of diseases in which the hemoglobin molecule's synthesis or structure is incorrect. It is passed down through the generations (inherited). Hemoglobin C disease, hemoglobin S-C illness, sickle cell anemia, and thalassemia are all examples of this group of diseases (National library of medicine, 2020).

In sickle cell disease occurs a mutant within a β-globin gene, which results in unusual structures in the hemoglobin β-globin chain. Whereas, thalassemia is characterized by improper synthesis and production of the α- or β- globin, which results in a decrease or absence of globin. Patients are classed as alpha or beta thalassemia depending on the location of the defective gene (Alanazi, 2020).

2.5. Classification of Hemoglobinopathic Disorders

Haemoglobinopathies are the most frequent recessive monogenic illnesses in the world. They can be divided into two categories: thalassemia syndromes and structural hemoglobin variations (abnormal hemoglobin). α, β, and δβ thalassemia are the most common types of thalassemia with clinically importance. structural hemoglobin variants; HbS, HbE, HbC, and HbD are the most common and clinically important structural hemoglobin variants. The cost of treating patients is a significant part of the worldwide disease burden (Sanctis *et al*., 2017).

2.5.1. Thalassemia Syndromes

Thalassemia syndromes are autosomal recessive hemoglobinopathies that are caused by a decrease in the rate of synthesis of one or more of the hemoglobin polypeptides chains. The severity of this disease's clinical spectrum can range from asymptomatic test abnormalities to intra uterine death(Lulla *et al*., 2020).

Thalassemias are a series of recessively inherited illnesses characterized by low or no hemoglobin production and varied degrees of chronic anemia. The high incidence of thalassemia in the area stretching from Mediterranean, Sub-Saharan Africa, and the Middle East, basin to Southeast Asia is explained by the evolutionary link between the thalassemia carrier state and malaria resistance. Thalassemia has also been introduced to Europe and the Americas as a result of population migrations, where the disease was formerly quite infrequent (Taher *et al*., 2021).

It considered as a severe public health issue that affects people all over the world, especially in underdeveloped nations. There are several types of this disorder. Two of these variants, known as alpha and beta are the most prevalent. Furthermore, hemoglobin which carries oxygen to all of the cells in the human body, is affected in any of these two types of thalassemia. According to studies, the spread of beta thalassemia major is a big threat to middle- and low-income countries all over the world. Parents and children are subjected to a significant psychosocial burden as a result of the disease (Ul Hassan Rashid *et al*., 2020).

B-Thalassemia's are autosomal recessive hereditary illnesses characterized by decreased adult hemoglobin (Hb) production due to decreased b-globin chain synthesis, leading in defective erythropoiesis. β-Thalassemia major (β-TM) is a severe variant of b-thalassemia that requires blood transfusions (Yassouf *et al*., 2019).

A deficiency in beta-globin protein production is usually the cause of beta-thalassemia, which is the most common kind of thalassemia. The three types of β thalassemia are mild thalassemia, intermediate thalassemia, and major thalassemia. In the first two categories, a single beta-globin gene fails to result in a 50% reduction in beta-globin protein in the cell. The absence of β-globin chain production in thalassemia major, often known as Cooley anemia, is a transfusion-dependent clinical feature of thalassemia (Shareef & Obaid, 2017).

The quality of life among children with genetic blood diseases can be greatly affected by chronic low hemoglobin level, as well as the continuous exposure to blood transfusion procedures, in order to avoid early and late complications such as spleen enlargement, hepatomegaly, growth retardation and heart failure. It exposes the child to stresses that increase the burden of the disease on the child, family and community itself, therefore the procedures standard must be used to improve their quality of life and the possibility of complications (Lulla *et al*., 2020).

2.5.1.2 Type of thalassemia

α- and β-thalassemia are the most frequent types of thalassemia, the most important genetic variation of thalassemia is β-thalassemia which is occur due to impaired production of β-globin chain type, lead to severe transfusion dependent anemia which significantly leads to poor quality of life (Aljeesh, 2016).

Thalassemia knew as a genetic autosomal recessive hematologic disease wherein the synthesis of one of the globin subunits of the hemoglobin is reduced or absent. Based on the kind of globin chain containing two main types α- and β -thalassemia can be recognized. In addition, other types of thalassemia can result from imbalance production of 2 to 4 different globin chains (δβ-, γδβ-, and εγδβ-thalassemia) (Maatook *et al*., 2015).

Thalassemia trait as the mild type occurred when one of a child's globin genes is defective and be inherited just from a single parent. People who are affected with minor thalassemia oftentimes don't have available symptoms and can live their life normally without management. Also, individuals with a thalassemia minor are usually known as thalassemia carriers. As an example, if a person has major α-thalassemia, he or she has (2) abnormal α-globin genes transmitted from both parents. Whereas, if a person was a β-thalassemia carrier, he or she inherited (1) abnormal β-globin gene from only a single parent (Kim & Tridane, 2017).

The second as the moderate one; β-thalassemia intermedia , which is less severe than those of β-thalassemia major. Early childhood or later in life, signs and symptoms occur. Affected people have mild to moderate hypochromic microcytic anemia with no visible clinical symptoms. The difference between major and intermedia is that (Major) requires chronic red blood cell transfusions whereas (Intermedia) requires no or infrequent transfusions (Pedullà *et al*., 2015).

Anemia with moderate hemolysis and the capacity to conserve a level of hemoglobin more than 7 g/dL are symptoms of beta-thalassemia intermedia. Because of the heterogeneity of the phenotypic, serious instances of intermediate thalassemia can manifest early in childhood two– six years, whereas lesser serious variants can manifest later in life with mild anemia and other problems (Sayani & Kwiatkowski, 2015).

The severe one is the major thalassemia (Homozygous β-Thalassemia) Cooley anemia or Mediterranean anemia is another name for thalassemia major. A blood spot test is used to diagnose it at birth. Because this is a β-chain hemoglobin deficiency, symptoms do not appear until the HbF had been fully exchanged by HbA in the second half of a baby's first year of their lives (Pillitteri, 2018).

β-TM is occurred by complete absent of β globin formation due to several copies of HBB gene mutation. This kind of β-thalassemia is severe and can appear within the first 2 years of life with clinical manifestation such as severe anemia, growth retardation, jaundice, and distended organs, should be adopted a regular blood transfusion in the treatment of β-thalassemia major to supply them with enough volume of mature RBC (Alotibi, *et al*., 2019).

2.5.2. Structural Hemoglobinopathy

Gene abnormalities that affect the structure of hemoglobin cause structural hemoglobin variants, such as those found in sickle cell disease, hemoglobin C (HbC) illness, and hemoglobin E (HbE) disease (Lee *et al*., 2019).

2.5.2.1. Sickle Cell Disease

SCD is a term that refers to a range of genetic illnesses that affects millions of individuals around the world and all linked to the existence of HgbS. Although SCD is occasionally used interchangeably with SCA, this is incorrect. HgbSS disease and homozygous sickle cell disease are the correct terminology for SCA (Hockenberry *et al*., 2019). The affected child receives two defective hemoglobin genes from both parents, making it an auto-recessive blood disorder. In SCD, abnormal hemoglobin prevents the formation of red blood cells. This causes the normally round red blood cell to become elongated and crescent-shaped, resulting in a reduction in red blood cell movement via blood channels to various regions of the body (Nwanonyiri *et al*., 2019).

The chronicity of SCD. Present the recurrent pain crises, persistent anemia, and other debilitating problems. One of the most serious consequences of the problem is impaired cognitive function. When compared to healthy peers and normative samples, children with SCD have cognitive deficiencies in multiple domains (Yarboi *et al*., 2017).

HbSS occurs when persons inherited one mutated sickle gene (S) from every parent. This formation is frequently referred to as SCA and is the more prevalent form of SCD. HbSC is a milder type, which occurs when a sickle gene (S) is transmitted by one parent and an aberrant HbC gene is transmitted by another parent (Kaufman *et al*., 2018).

Sickle thalassemia disease is caused by a mix of sickle cell and β-thalassemia traits. Some normal adult hemoglobin can still be formed in the β+ (beta plus) form. There is no ability to make normal adult hemoglobin in the β0 (beta zero) form. While, HbS beta-thalassemia (HbSβ+, and HbSβ0) occur when one sickle gene is transmitted by one parent and one β-thalassemia gene is an inheritance from another parent. Beta-thalassemia causes inadequate hemoglobin production and a result is a deficit of hemoglobin in the RBC (Kaufman *et al*., 2018; Hockenberry *et al*., 2019).

Accordingly Homozygotes (HbSS) suffer from sickle cell anemia (SCA), which leads to acute and chronic complications including vaso-occlusive crisis, acute chest crisis and hemolytic crisis. SCA clinically behaves as a multi-genic trait with exceptional phenotypic variability (Piel , 2015).

2.5.2.2. Hemoglobin E

HbE is a type of defective hemoglobin caused by such a qualitative mutant in the β-globin gene, and it is the world's 2nd most frequent globin mutant. Patients can affect by silent hemoglobin E trait (HbAE) or benign homozygosity hemoglobin E illness (HbEE). Depending on the β-thalassemia mutation, compound heterozygote HbE thalassemia causes symptoms varying from mild to severe anemia. HbE/β-thalassemia is virtually exclusively seen in people of Southeast Asian heritage in California, with an incidence of 1: 2,600 births (kliegman, 2020).

2.5.2.3 Hemoglobin C

The HbC mutation occurs at the same location as HbS, but instead of valine, lysine is substituted for glutamine. Hemoglobin C trait (HbAC) affects 1 in 40 African Americans, and homozygous hemoglobin C illness (HbCC) affects 1 in 5,000. HbAC has no symptoms. HbCC can cause mild anemia, splenomegaly, and cholelithiasis, as well as rare cases of spontaneous splenic rupture. There is no splenic dysfunction. Typically, this disease is detected through neonatal screening programs. The red cell membrane is disrupted by HbC crystallization, and hemoglobin C crystals were probably observed on marginal smears (kliegman, 2020).

2.5.2.4 Hemoglobin D

At least there 16 different types of HbD exist. HbD-Punjab (Los Angeles) is an uncommon Hb found in just 1-three percent of West Indians and a few European of Asian descent. When combined with HbS, it causes sickle cell disease symptoms. Heterozygous HbD, also known as hemoglobin D trait (HbAD), has no symptoms. Hemoglobin D disease (HbDD) is a mild to severe anemia with splenomegaly caused by homozygous hemoglobin D mutations (kliegman, 2020).

2.6.Etiology

Hemoglobin is a tetramer made up of two globin chains in pairs. Hemoglobinopathies are diseases caused by abnormalities in these proteins (Kliegman, 2020).

The inherited genetic hemoglobin disorders are controlled by a single gene that passed down from parents to children from one generation to the next. All types of hemoglobinopathy are inherited in the same manner. For example, Parents carrying mutated thalassemia gene be able to pass it to their child. A child who inheriting one mutated gene is mean carrier, this type of disorder considers to be a thalassemia trait. Most carriers have a normal healthy life, if both parents have TMT there are 25% probability rate in every pregnancy that their child will have β thalassemia major, a 50 % possibility rate that a child will have TMT and a 25% possibility rate that a child will be without mutation (Abdulsattar& Hattab,2017).

Furthermore illustration individuals who inherit only one altered allele (also known as traits) are said to be heterozygous, but those who inherit two affected alleles are said to be homozygous, which usually results in a more severe phenotype (e.g., severe anemia) (Pei *et al*., 2021).

2.7. Pathophysiology

SCD is an inherited illness that impairs the structure of hemoglobin, the oxygen-carrying protein in red blood cells. A pathogenic mutant in the beta-globin (HBB) gene changes the structure of hemoglobin, causing hemoglobin to polymerize and deform red blood cells into a sickle or crescent shape (Grygiel *et al*., 2021).

Normal RBCs are spherical, soft, and elastic, and can survive up to 120 days, sickle RBCs are rigid and take on a (C) form in deoxygenation due to a hemoglobin mutation. Because of their odd shape, these cells barely living for 10–20 days and have trouble migrating successfully through blood channels. The sickled shape restricts RBC mobility through blood vessels, contributing to a variety of medical issues associated with SCD, such as extreme pain and end organ damage (Kaufman *et al*., 2018).

Thalassemia is an inherited hemolytic blood disease characterized by the reducing the production or lack synthesis of one or more hemoglobin chains. Hemolysis occurs as a result of the problem, resulting in a shortened RBC life span (Kweka *et al*., 2020). In a healthy person, there is a coordination in the synthesis of the α- and β-globin chains, this chain found in the normal hemoglobin for α2β2 (HbA) will be formed. While in thalassemic person there is a defective synthesis of either α- or the β-globin chain (Hisam *et al*., 2018).

β-thalassemia is caused by an imbalance in the α-/non α-globin chains. α-Globin tetramers aggregate and precipitate in erythroid precursors, generating inclusion bodies that induce oxidative membrane deterioration and severe early death of RBC precursor in bone marrow by apoptosis (ineffectual production of RBC). Hemolysis has a minor role. Extramedullary erythropoietic tissue masses can occur from erythroid marrow hypertrophy in the medullary and extramedullary regions causing abnormalities of the skull and face and can cause cortex weakening and pathologic breaking of long bones. The lipids membrane component of defective RBC can cause thrombosis problems, particularly in patients who have had their spleen removed (Origa,2017).

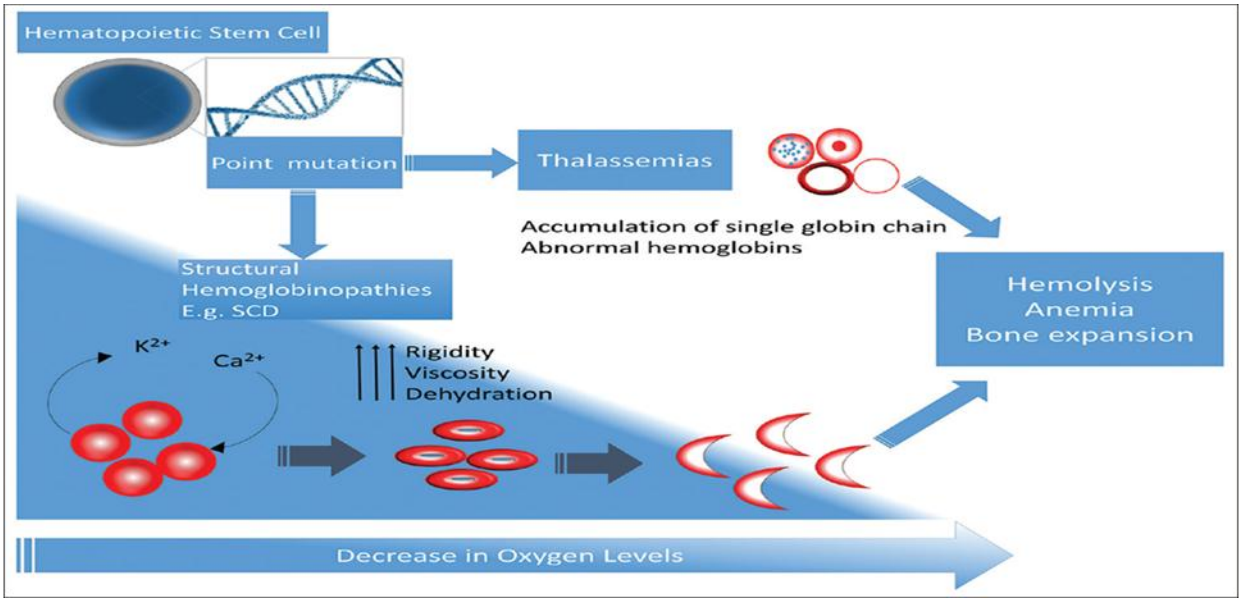

Figure (2-2) Point mutation within hematopoietic stem cells may lead to structural hemoglobinopathies or thalassemia. The cellular phenotypes are more prominent when oxygen level is decreased (Al Alwan *et al*.,2020).

2.8. Clinical Manifestation

Thalassemia variants and main defective hemoglobin's interact to create a broad range of clinical manifestation that varying in severity (Sanctis *et al*., 2017).

β-thalassemia have a wide range of clinical symptoms, that range from asymptomatic occurrence with minor (silent) mutations to mild hypochromic anemia to moderate and severe lifelong transfusion-dependent anemia with multi-organ dysfunction (Sanctis *et al*.,2017).

Within the first two years of life, signs and symptoms of β-Thalassemia major become visible. Affected people may have an enlarged spleen, liver, or heart, as well as deformed bones (Pedulla *et al*.,2015).

Anemia, growth retardation, and skeletal abnormalities are common in TM patients. It's challenging to stick to treatment plans that entail regular blood transfusions and iron chelation treatments. Multiple transfusions can result in high levels of iron in the body, which can lead to endocrine problems (like diabetes mellitus and hypogonadotropic hypogonadism), growth retardation, hepatic fibrosis, cardiac dysfunction, colored skin, and psychosocial issues like depression and executive function deficits (Örengül *et al*., 2019).

Hemoglobin S polymerizes upon deoxygenation, resulting in a phenotype marked by a chronic hemolytic process, which is often exacerbated by a variety of crises, particularly vaso occlusive ones, and leads to multi-organ damage. In addition to its physical effects, SCD has a social, psychological, emotional, and academic impact on the affected individual's life (Ameer Tahir *et al*., 2019).

Vaso occlusive crises are the disease's first major clinical manifestation. In SCD, tissue remodeling and organ dysfunction are thought to be caused by repeated VOC-related ischemia assaults and subsequent reperfusions. As a result, ischemia and reperfusion may have a role in SCD morbidity and death. Chronic hemolytic anemia is a second prominent symptom of the condition, which is frequently resulting in severe hemolysis that erythropoiesis does not counteract. This results in extremely reduce hemoglobin and hematocrit levels usually “6–8 g/dL and 25%–30%, respectively”. Patients with SCD also have desaturation of the arteries and collectively decrease hematocrit and hemoglobin concentration, desaturation of the arteries and decrease p_{50} produce hypoxemia that may lead to poor delivery of O_2 to tissues (Merlet *et al*., 2019).

Sickle Cell Anemia (SCA), the most prevalent and severe form of SCD which lead to recurrent vaso-occlusion events produce endothelial dysfunction and inflammation, resulting in severe seizures and increasing tissue and organ damage. These diseases cause major problems in children as early as childhood, including stroke and vasculopathy, as well as acute thoracic syndrome or vaso occlusive crisis in younger children (Brito *et al*., 2020).

Children with the current problem suffer from broad spectrum of symptoms and complications like acute chest syndrome, frequent painful episode, pulmonary hypertension and osteomyelitis. Because of their heightened susceptibility to significant morbidities such renal, respiratory, and cardiac problems, as well as stroke, they live shorter lives than the general population. In high-income countries, the median life expectancy is 40–60 years, whereas in low-income ones, it is substantially lower (Alhomoud *et al*., 2018).

2.9. Diagnoses of Hemoglobinopathies

Diagnosis of hemoglobinopathic disorders dependent on various methods including clinical and family history, complete blood counts (CBC), red cell indice, HbA2, HbF rates, sickling test, and Hb electrophoresis, high performance liquid chromatography and capillary electrophoresis (Shiva *et al*., 2017).

The first step in diagnosing thalassemia/hemoglobinopathy in anemic patients is to suspect the condition based on phenotype, family history, and appropriate laboratory screening test results. The diagnosis is established through molecular genetic confirmation by finding pathogenic mutations. The genetic variability of the disease and mutations overlooked by traditional sequence studies made molecular genetic diagnosis difficult in the past (Lee *et al*., 2019).

2.10. Treatment and Management

The typical treatment choice is dependent on the recognition and molecular characterization of hemoglobinopathies patients a necessary condition for selecting suitable treatment. Other factors that are considered important for patient stratification include clinical manifestation which can extant from none to serious grade (Karamperis *et al*., 2021).

Clinical manifestation and optimal therapeutic methods vary for patients with TM and SCD. Nonetheless, RBC transfusion is a crucial part of both disorders' management. Patients with TM need RBC transfusions for the rest of their lives, but those with SCD only need transfusions in specific clinical conditions (Al-Riyami & Daar,2018).

The survival of individuals with SCD has improved as a result of a series of treatment-oriented efforts, which have reduced morbidity and mortality while also increasing life expectancy. Prophylactic use of penicillin in children under the age of five, as well as access to particular immunobiological and drugs for routine use, such as folic acid and hydroxyurea, are examples of therapeutic improvements. Blood transfusions are recommended when children do not respond to pharmaceutical treatment (Gesteira *et al*., 2020).

Poor drug adherence raises disease burden, raises healthcare costs, and expands disparities in health. Health-care utilization, treatment success rates, missed school/work, and symptom improvement have all been used to assess treatment adherence (Smaldone *et al*., 2019).

2.10.1. Transfusion and Iron Chelation Therapy

Transfusion therapy has two main goals: providing normal erythrocytes to reduce chance of anemia and reducing inefficient erythropoiesis, effectively managing the downstream pathophysiological mechanisms in thalassemia (Lulla *et al*., 2020).

The requirement for blood transfusions is determined partially by the severity of anemia and, more crucially, the failure to adjust to anemia, which manifests itself in a poor development or nutrition, rised cardiac output, tachycardia, or rising symptoms of inappropriate erythropoiesis (deformities of bone, and hyper splenomegaly) (Sayani & Kwiatkowski *et al*.,2015).

In transfusion-dependent patients, regular transfusions are given to achieve pretransfusion hemoglobin levels of 9 to 10.5 g per deciliter (11 to 12 g per deciliter in patients with heart disease). Although transfusion techniques vary by area, improvements in donor blood screening and preparation have reduced the prevalence of alloimmunization and blood borne diseases in most countries (Taher *et al*., 2021).

Chronic transfusion therapy includes significant problems such as acute life-threatening complications (bacterial infection, anaphylaxis, and acute hemolytic reaction), infection, and iron overload which can cause serious multi system organ damage (Algiraigri *et al*.,2018). SCD alloimmunization is a significant side effect of blood transfusions, and it frequently results in life-threatening, acute and delayed transfusion responses (Alkindi *et al*., 2021).

In Cooley's anemia patients who get regular transfusions, hemochromatosis is unavoidable because of human body lack a way to eliminate extra iron. Iron overload is poisonous to multiple tissues resulting in cardiac failure, liver cirrhosis, growth retardation, and a variety of endocrinal disorders (Lulla *et al*., 2020).

Children with SCD, iron deposits primarily in the liver, with less in the heart and endocrine organs (Alkindi *et al*., 2021).

Serum ferritin measures and hepatic and myocardial MRI should be used to check for iron overload in transfusion-dependent patients, and iron chelation should be given promptly after a transfusion of 10 packed red-cell units or when the serum ferritin level is 1000 ng per milliliter or above (Taher *et al*.,2021).

Deferoxamine, deferiprone, and deferasirox are three iron-chelating medicines now in use and licensed by the US-FDA (Shareef & Obaid, 2017).

There is evidence that iron chelating medications minimize tissue damage and enhance life expectancy in people who require long-term iron chelation therapy. The goals of iron chelating therapy in these patients are to reduce iron burden, reduce risk of tissue damage, especially in specific key organs like the heart and liver, improve life survival, provide 24-hour protection against toxic effects of iron like Labile Plasma Iron, and finally, decrease the gap free of iron chelating drugs (Al-Madeni *et al*.,2018).

2.10.2. Hematopoietic Stem Cell Transplantation/ Gene Therapy

The only available curative therapy for hemoglobinopathies including SCA and thalassemia is HSCT, or gene therapy, but is only available to a few numbers of patients with compatible donors (Al Alwan *et al*., 2020; Karamperis *et al*., 2021; Musallam *et al*., 2021; Sinha *et al*., 2021).

Significant advancements in medical treatment of problems related to transplants have led to a greater number of individuals being considered eligible for transplantation. unluckily, the patient percentage who on a higher success rate doesn't reach 10%, therefore access is still restricted. Major predominant limitations include a lack of appropriate donors and the increased likelihood of immunological adverse effects. The matching of HLA between both donors, and recipients determines if, a transplant may be conducted successfully. Other considerations, such as the patient's clinical state (comorbidity) and age, are also taken into account as additional constraints, resulting in a great variety of success rates among patients (Karamperis *et al*., 2021).

In young patients who have a Human Leukocytes antigen matched donor, bone marrow transplantation (BMT) may offer the chance of a cure. The rates of success of the BMT were in the 80–90% range, whereas the mortality rate was around 3%. There are no randomized controlled trials (RCTs) available. which have investigated the safety and effectiveness of non-identical donor bone marrow transplantation in β-thalassemia patients who require blood transfusions. If there isn't an HLA-matched compatible donor for this person. Bone marrow transplantation is another option (BMT) (mismatched donor) from a haploidentical mother to her child be put to use in a study of 31 participants, the thalassemia-free group came out on top. The survival rate is 70%, rejection is 23%, and death is 7%. The most effective outcomes are for extremely young people (Elsayed &Abd El-Gawad, 2015).

The majority of children with the disease die due to heart failure in the adolescent period or late adolescence if they didn't receive HSCT (Pillitteri, 2018).

2.10.3. Splenectomy

Splenectomy may be essential to decrease discomforts and also to decrease hemolysis rates of erythrocytes and the number of blood transfusions required (Pillitteri, 2018).

As evidenced by the prevalence of splenomegaly due to excessive destruction of defective RBCs, extramedullary hematopoiesis, and transfusional overload in thalassemia, the spleen is the most usually damaged organ. Splenomegaly increases the need for blood transfusions even more. When hypersplenism increases the need for blood transfusions and hinders proper control of body iron with chelating treatment, splenectomy is recommended in transfusion-dependent patients. An enlarged spleen without an increase in transfusion requirements isn't always a sign that surgery is needed. Patients with hypersplenism may have splenomegaly ranging from mild to severe, as well as neutropenia or thrombocytopenia (Saleh *et al*.,2018).

2.11. Nursing Diagnosis

- “Ineffective tissue perfusion related to generalized infarcts due to sickling”.
- “Compromised family coping related to long-term care needs of child with a chronic hematologic disorder’’.
- “Risk for situational low self-esteem related to changed physical appearance” (Pillitteri., 2018).

2.12. Nursing Interventions

The nurse plays essential role in providing care for patients with chronic disease including hemoglobinopathic disorders like thalassemia and sickle cell anemia (Chouhan & Pujari,2021).

Patients with hematologic disorders as other patients with chronic and disabling illnesses, require continuous lifelong treatment and face a variety of mental health issues, as well as social and economic issues, all of which interfere with treatment and follow-up strategies in some way (Shahraki-Vahed *et al*.,2017).

The important nursing intervention for patient with ineffective tissue perfusion including O_2 administration by either nasal cannula or mask if ABG shows a decreased level of PO_2. Carefully monitoring the flow rate of oxygen, and using pulse oximetry to evaluate level of saturation. As well as promote bed rest to minimize O_2 expenditure (Pillitteri., 2018).

Family members are impacted by one another, and a family member's disease has a direct impact on the entire family. Stress placed on one family member will have an impact on the entire family. Their energy levels are depleted, putting them at danger of physical, mental, and social isolation, as well as despair, frustration, helplessness, dread, shame, and a desire to die. As a result, these patients are often known as "hidden patients" (Shahraki-Vahed *etal*.,2017).

Nursing staff makes the first contact with the health-care team, thus it's critical that they have great communication skills. Empathy and sensitivity are crucial qualities for a nurse to have when managing a patient. Clear communication allows the patient to convey his or her concerns and fears while also allowing the nurse to offer support, encouragement, and reassurance, resulting in positive feedback (Azize *et al*.,2015).

The nurse is a crucial component in the effective management of hemoglobinopathies. Specialists of nursing afford knowledgeable, skilled sustenance and reinforcement all over and often standardize treatment establishments. The pediatric nurse is in a unique position to create an obligatory relations between the haematologist, the patient, as well as other healthcare professionals and necessary services since she knows the patient, family, and societal situation intimately. The nurses deliver the best possible care (Elsayed &Abd El-Gawad, 2015).

Because some adolescents' sexual maturity is delayed, both children and parents may require consultation to accept this; nevertheless, they can rest confident that once puberty changes do come, they are sufficient and only emerge later than typical. children may require support and encouragement reinforcement during their growing years, to promote their self-respect as they learn to cope with the effects of chronic hematological problems (Pillitteri., 2018).

By recognizing the key SCD-related problems and providing patients and their caretakers with relevant educational material, nurses who care for patients with SCD have the chance to enhance health outcomes and quality of life for these patients. The authors detail the primarily related health consequences as well as the genetic, hematologic, and clinical aspects of sickle cell disease (Tanabe *et al*.,2019).

2.13. Complications

The β-TM phenotypic is a severe condition that requires lifelong transfusions and has plenty of complications. Hypothyroidism, hypoparathyroidism, hypogonadism, growth retardation, diabetes mellitus, heart malfunction, and liver illness are examples of the latter. The frequency of these issues varies significantly among β-TM cohorts and is linked to the quality of management and follow-up offered to these patients, with industrialized countries having a significantly higher quality of life and fewer complications than developing countries (Sadullah *et al*.,2020).

Extramedullary hematopoiesis (EMH) is a common complication of erythroid marrow hypertrophy. Its symptoms include bone and facial abnormalities, osteoporosis with pathological long bone fractures, erythropoietic tumors mostly affecting the spleen, liver, lymph nodes, chest, and spine, and a tendency for thrombotic problems. The spleen's size is partly a result of its important role in removing damaged red cells from the bloodstream. Leg ulcers are very common. In thalassemia intermedia, cardiac involvement is recognized by an increased output status and pulmonary hypertension, with typically preserved systolic left ventricular function Heart muscle siderosis is a relatively uncommon condition (Cappellini *et al*.,2021).

In children and adolescents with homozygous β-thalassemia, growth retardation is common. After four years, longitudinal growth patterns show rates that are slower than normal controls. After the pubertal growing spurt fails, bone age is usually delayed after the age of 6-7 years, and growth retardation becomes noticeably severe (Soliman *et al*., 2018).

Kids with SCD are frequently thinner and shorter than healthy peers throughout childhood and early adolescence. The incidence of low body weight in American children with Sickle cell disease is (41 percent) and (25 percent) with severe undernourishment, with a wasting prevalence of (11%). Multiple factors appear to influence growth failure in SCD, including hematological and circulatory state, endocrine and metabolism functioning, and dietary state (Mandese *et al*., 2016).

People with SCD are extremely vulnerable to infection, particularly respiratory tract infections and septicemia. The genotypes HBSS and (Sβ0Thal) are the most severe in general. Functional hyposplenism is frequent in children with homozygous SCD; it affects 90% of children under the age of five and is linked to vulnerability to encapsulated bacteria infection. Due to the disease severity being similar to homozygous SCD, children with the genotype Sβ0Thal are likewise prone to lose splenic function early in life. Infections are the primary cause of death among children with SCD and can begin as early as infancy. This is mainly related to splenic dysfunctions, which cause immune system weakness (Rankine-Mullings & Owusu-Ofori,2021).

Persons with SCD have an increased risk of acute lung disease and viral infections when compared with the general population. according to hospital records, kids with SCD are approximately 56 times more likely than children without SCD to be hospitalized in the US in 4 states for two influenza seasons (Chowdhury & Anwar, 2020).

Chronic pain is other complication for sickle cell disease, which affects adolescents and adults more than children. This agony is distinct from crisis pain, and many people find it difficult to describe. Chronic pain can be mild or severe, and its cause is unknown. SCD sufferers are also prone to anemia, or a low RBC count. Although severe anemia, which is life-threatening , may occur in this patients , the severity levels are often mild to moderate. SOB, lethargy, vertigo and pale complexion are all symptoms of severe anemia. Children with SCD may experience reduced growth rates due to anemia (Sabet, 2019).

2.14. Prognosis

Early detection of hereditary hemoglobinopathies is critical in order to provide proper care to people who are at risk and avoid worsening of the disease and associated comorbidities (Pei *et al*., 2021).

Many studies show that if SCD is diagnosed early, ideally within the first three to six months of life, life-threatening early problems can be generally avoided. The introduction of NBS for SCD has resulted in a considerable reduction in morbidity and mortality among those affected. Since the establishment of standardized NBS for SCD in the United States, the death rate in affected children aged 1 to 4 years has fallen by 50%, and males' life expectancy has increased from 14.3 years to 42 to 53 years, and females' life expectancy has risen from 46 to 58.5 years (Zhou *et al*.,2021).

Major thalassemia causes severe anemia in babies during the 1st year of their life, whereas intermediate thalassemia causes asymptomatic mild anemia. People with TM dieing in their childhood and teenage years in developing and undeveloped nations due to a lack of public access to virus-tested blood transfusions and timely iron removal from the body. Iron overload and excess serum ferritin are the underlying causes of death in these patients, leading to heart illness, hepatic and endocrine diseases, infections, clotting , anaemia, malignancies, and dieing finally (Ansari-moghaddam *et al*., 2018).

Hemosiderosis and tissue dysfunction, including hepatic, cardiac, and endocrine problems, can result from frequent blood transfusions. These patients die in their second decade of life as a result of the issues mentioned, but today the survival rate has great improvement related to advance in therapies, especially after using the chelation therapy (Badfar *et al*., 2017).

2.15. Preventive Measures for Hemoglobinopathic Disorders

Multiple recommendations were published by the American Faculty of Obstetricians and Gynecologists for families at high risk of having a child with hemoglobinopathies, including increased genetic counseling and education about hemoglobinopathies, their risk, and illness management. Most importantly, PND and PGD medical technologies are a significant tool for high-risk families who want to ensure that their child is free of hemoglobinopathy (Alanazi, 2020).

The detection and counseling of carriers (heterozygous) to discourage marriage between carriers is the foundation of a main preventative program. Premarital screening (PMS) for thalassemia and other genetic illnesses that can be prevented is common in many parts of the world. PMS is yet to be established in countries with a high proportion of consanguineous marriage (40%), with more than 85 % of those marriages being between first cousins and traditional weddings, which could make acceptance difficult. As a result, PMS for hereditary disorders is an essential technique for reducing the social, emotional, and financial burden on families and society in high-risk populations. PMS has proven to be effective in many parts of the world (Al-Kherbash *et al*.,2017).

2.16. Characteristics of Adolescents

Adolescence is a complex stage of life marked by physical and psychosocial development, enhanced cognitive capacities, and the formation of personal and social identities (Lucio *et al*., 2019).

According to (WHO, 2019) Adolescents are those between the ages of 10 and 19 year, whereas youth are those between the ages of 15 and 24. Young People' refer to anyone between 10 and 24 years old.

Adolescence is a phase of rapid growth and maturation in humans. During this time, a spurt in somatic growth and the onset of secondary sexual characteristics, such as menarche and spermarche, occurs. This is also a time when nutritional needs will be higher. Adolescence represents for around 20% of total stature growth and up to 40% to 50% of body weight gain as somatic growth (Patil *et al*., 2013).

Important physical, sexual, emotional, and social developmental changes occur continuously as a person grows from childhood to maturity. This transformation, although creating growth opportunities, also poses a risk to their health and well-being. Despite the popular notion that adolescents are a healthier age group, they confront a variety of public health issues (WHO,2019).

2.16.1. Physical Wellness for Adolescents

Physical wellness entails paying attention to and caring for adolescents body in order to achieve optimal health and performance (University of New Hampshire,2022).

Self-management is a collection of tasks that persons with chronic illnesses must implement into their daily routine in order to improve their health (Smaldone *et al*.,2019).

Adolescents with SCD confront a unique set of challenges. Due to their sensitivity, factors including stigma, delayed menarche, and delayed physical development may have a detrimental impact on them, leading to a negative self-image and dangerous behavior (Forrester *et al*.,2015).

Muscle dysfunction compromise's physical capacity, which is already compromised by anemia, cardio-respiratory restrictions, and hemoglobinopathic disorders. Muscle dysfunction affects daily life physical tasks in addition to physical fitness. Several studies have found that SCD patients have lower physical activity energy expenditure than healthy participants. Furthermore, the risks of acidosis, hyperthermia, dehydration, oxidative stress and other side effects of excessive physical exercise may cause parents and/or physicians to prevent children and adolescents with SCD from participating in physical activities., resulting in SCD patients adopting hyper sedentary lifestyles. Hyper sedentary behavior exacerbates physical deconditioning, particularly muscle deconditioning, trapping SCD patients in a vicious cycle that results in decreased muscle function, physical fitness, and movement (Merlet *et al*., 2019).

Major beta thalassemia has a severe impact on physical health, resulting in physical deformity, growth retardation, and puberty delays. It has a negative impact on physical appearance, such as bone abnormalities and small stature, which contributes to low self-esteem. When children reach school age, they want independence and become more conscious of the differences between themselves and others, the impact of disease on many aspects of life becomes more apparent (Abdulsattar & Hattab, 2017).

Children see differences between themselves and others that are related to either their physical appearance, growth failure, or bone deformities, or their inability, such as a loss of energy to complete daily responsibilities and prior physical activities that they used to prefer (Al Ebrahimy *et al*., 2016).

Hemoglobinopathic patients face obstacles in school, obtaining work, and/or getting married as a result of the disease's functional and physical limits. As a result, persons with thalassemia are likely to have a lesser quality of life than those who do not have the condition (Töret *et al*.,2019).

Fatigue has primarily been studied in children's populations as a symptom that impairs their quality of life. Despite the fact that fatigue has been found as a primary symptom affecting both health and quality of life in people with sickle cell disease, it has also been identified as a major symptom influencing quality of life in patients with thalassemia major (Aslani *et al*.,2018).

Identity formation is an important element of teenage development, and it entails incorporating any chronic disease into one's body image and self. This is a gradual process, and adolescence is a time when fresh questions about the source, nature, and consequences of any sickness or impairment arise. Information and education on these chronic illnesses must be provided as soon as possible and in an appropriate manner (Abdulsattar & Hattab, 2017).

Chronic disease usually affects the progress of growth and development. Treatment requirements, recurrent hospitalization and surgery when necessary lead to rising worries about physical aspect, effect on the process of winning independence and a healthy relationship with family and friends. Also, developmental problems interfere the children and adolescents' ability of being responsible for managing their disease (Elzaree *et al*., 2018).

Pain affects movement and the capacity to perform everyday tasks, such as walking, showering, and doing chores. The term "activities of daily living" refer to all of these actions (ADLs). Physical (e.g., clothing, hygiene, grooming, feeding), cognitive (e.g., remembering of taking prescriptions, playing a game), complicated ADLs (marketing, cooking), and combinations of ADLs (which combine all categories of ADLs) are all examples of ADLs. Adding to the proven deterioration in ADLs linked to chronic ache, studies has also demonstrated that ache is linked to weaken ADLs in a variety of other chronic conditions, including cancer, chronic low back ache, fibromyalgia, and chronic ache problems in particular. There are only a little amount of data that shows decreased ADLs in particular affected with SCD. but there are several studies for people with SCD demonstrated that pain is linked with low physical performance (Kaufman *et al*.,2018).

2.16.2. Psychosocial Wellness for Adolescents

Adolescents with chronic illnesses' emotional health might be influenced by family connections and psychosocial concerns. Family bonds have a stronger impact on the well-being of these teenagers than the sickness itself. Male gender, psychiatric disease or crime in a parent, and low socioeconomic level are all psychosocial factors linked to poor emotional health in these teenagers. The duration of the sickness, its visibility, how it limits mobility, and the patients' predicted survival are all factors that influence emotional well-being. Positive temperature, above-average IQ, social competency, a positive relationship with at least one parent, family closeness, and adequate rule establishing by parents are all a protective factor(Abdulsattar & Hattab, 2017).

Emotional and behavioral disorders are one of various issues that might obstruct a child's development, including psychosocial issues, childhood depression, and other mental diseases. Children's emotional and behavioral disorders are a critical issue since they impair their development and reduce productivity and quality of life (Tawarina & Diba,2020).

The promotion of mental health well-being among the world's teenage populations is crucial for public health and social well-being. This is especially true for teenagers with chronic illnesses, since research has shown that these groups are more likely to acquire psychological problems (Orth & Van Wyk,2020).

Patients and their families experience a significant psychological strain due to the chronic nature of disease and its intensive and demanding treatment. Psychological anguish is exacerbated by social isolation, low self-esteem, poor academic achievement, and stigmatization (Pranandita, 2021). Stress, anxiety, and depression are comorbid illnesses in sickle cell and thalassemia (Lyrakos *et al*., 2017).

Children with thalassemia have a considerable psychological impact, causing emotional problems, hopelessness, and social integration difficulties. Thalassemic patients also have a variety of physical and psychological symptoms. Because the disease effects on the patient's physical appearance and body image, patient experiencing from negative self-image and low self-esteem. The effects of thalassemia on one's physical well-being can include physical deformation, growth inhibition, and delayed maturation (Akter *et al*., 2020).

The majority of SCD patients who live into the adolescence period describe psychosocial issues that adapt predominantly hard. The sensation of embarrassment, hopelessness, anxiety, and loneliness are among the psychosocial health serious complications of SCD-related sexual dysfunction. Socially stigmatization is the phenomenon in which a person with a communally undesirable attribute is rejected by the community, which becomes a serious worry for chronically ill patients, such with SCD (Ocansey, 2018).

SCD causes limitations in doing typical everyday activities as well as completely engaging in activities with their family, colleagues, and those around them on a regular (and infrequent) basis. Coping with health complications and prognosis of SCD, decreased wellbeing, depressed mood, changes in mood, anxiousness, separation anxiety, isolation, sleeplessness, ability to perform daily and role limitations, psycho - social somatoform disorder related to neurobehavioral impairment, insomnia, and analgesic dependence are all common psychosocial considerations in SCD. In comparison to the general population, SCD patients have a lower health-related quality of life. Due to medical issues such as severe pain crises, this population may find it difficult to Regularly, attend social events, work, or school. Long-term functional and medical difficulties in children and adults contribute to anxiety and depression (Kaufman *et al*.,2018).

2.17. Studies Related

First Study: A study done by (Moyen *et al*.,2021) an analytical cross-sectional study “Psychological Experience of Children and Adolescents with Homozygous Sickle Cell Disease in Brazzaville” aimed to detect the psychological problems in patients with SCD and to determine the associated elements, their results were dropping in self-esteem, 76.1 percent anxiety, 29.9 percent depression, and 5.5 percent a negative impact on everyday life in all cases were identified. These were a significant adverse impact of 39.3 percent. Psychological issues were linked to advance years of age, sickness duration, school and maturity delays, usage of upper-level analgesics, the number of problems and hospitalizations, and the occurrence of the complications.

Second study: A descriptive study was conducted by (Akter *et al*., 2020) on Bangladesh children with thalassemia aim to identify their living experiences. This study discovered four key elements and eleven subthemes: 1) Children's comprehension of thalassemia: perceptions of children who are unable to do daily tasks, as well as educational implications 2) dissatisfying experiences and disease management: feelings about long-term treatment, adapting, and alteration of body image, 3) familial relationship: variances from siblings, parent attention, and burdens feel, and 4) social relationships: sharing with peers, incapacity to socialize.

Third study: In a certain comparative study conducted in Babil province (2020), the aim was to determine the psychosocial development of patients with thalassemia and healthy adolescents and to compare the researched variables of thalassemic and healthy teenagers, which resulted in significant differences between healthy and thalassemic teenagers in comparison to the overall psychosocial development at (0.001), and a significant difference among thalassemic and healthy teenagers in comparison to the overall psychosocial development at (0.001) (Saeed & Aldoori,2020).

Forth study: “School performance of children with sickle cell disease in Basra, Iraq” a title conducted by (Abid *et al*.,2019), which aimed to compare the academic achievement of school-age children with SCD, to healthy pupils of the same gender and age, and to determine the factors that can impact the school success of these children. They concluded that patients with SCD exhibited considerably lower school performance and cognitive skills than their peers, and school absenteeism and patient age hurt school achievement.

Fifth study: In a cross-sectional study conducted in sub-Saharan Africa by (Kambasu *et al*.,2019), aimed to assess health-related quality of life among adolescents with sickle cell disease, which concluded that As reported by children and caregivers, the pain was negatively related to both physical and psychosocial function, whereas receiving the Pneumococcal vaccine was favorably linked with both physical and psychosocial functioning.

Sixth study: A certain comparative study was conducted in the Kurdistan region by (Mikael & Al-Allawi, 2018) on 100 thalassemic and 100 healthy children and adolescents, aimed to determine the “health-related quality of life” among thalassemia-affected Kurdish children in Iraq and teenagers, as well as the factors that influence it, which resulted that thalassemia patients' HRQoL scores were considerably lower than healthy participants, with the lowest scores in physical functioning. especially for those receiving 6 or more transfusions per year, those with HCV, those having parents who are uneducated and those who are taking iron chelation pills all had significantly poorer HRQoL scores. Health related quality of life was inversely associated with age, frequency of transfusions, and serum ferritin, but positively associated with age at first transfusion and age at diagnosis, according to Pearson correlation.

CHAPTER THREE

METHODOLOGY

Chapter Three

Methodology

This chapter designates the methods which considered a means to achieve the objectives settled for the purpose of the study.

3.1. Design of Study

A quantitative, “cross-sectional” descriptive design is used for assessment of the physical and psychosocial wellness among adolescents with hemoglobinopathic disorders in the Babylon province from October 19th 2021 to June 8th2022.

3.2. Arrangements for Administration

Before data collection, formal administrative permits were required, and they were obtained for the study as shown in Appendix (A).

1. The first permission was obtained from the University of Babylon College of Nursing to the Higher Studies Committee as a proposal presented which consist of the major statement and its objectives.
2. The research approval was established from ethical committee / college of nursing that involve five advanced scientific titles in the college, it sent to the settings of sample collection which specified in the study plan.
3. An official permission, approval has been received from "Ministry of Health" /Babylon health directorate /Babil Teaching Hospital for Maternity and Children/ hereditary blood disease Center in Babylon province.
4. An informed consent taken from the parents or guardians and the adolescent himself. Participants were asked for verbal informed consent.

3.3. Setting of Study

The current research is being conducted in the Babylonian province at Hereditary Blood Disease Center/Babil Teaching Hospital for Maternity and Children as the only one of its kind in the Babylon governorate; it was established in 1996, as a ward in the Marjan medical city, then transferred to the Babil teaching hospital for maternity and children, and finally became a special semi center in 2003. The center has 4 wards first two wards for male and last two for female with a total capacity of 35 beds, 18 nurses, 6 hematological specialist physicians, one rotator, and 10 laboratory specialists, and 35-40 patients are seen every day. This center offers a variety of medical health services such as diagnosis and therapeutic care (blood transfusions and desferoxamine).

3.4 The Sample of the Study

The sample of the study was selected Purposively (non-probability), made up of 150 adolescents of both genders who were chosen from records of patients who had been medically diagnosed with beta thalassemia intermediate or major, sickle cell anemia, and sickle thalassemia. A total of 1323 patients from various age groups registered in the center, with 679 patients had thalassemia major, 259 patients had intermediate thalassemia, 44 of them had minor thalassemia, and 40 of them with alfa thalassemia. 6 patients had hemoglobin C, 3 patients have hemoglobin D, 83 with sickle thalassemia, 2 patients with delta thalassemia, and plastic anemia plus G6PD approximately 207 patients. There are 300 patients in the teen age group. The sample drawn as 50% from a total of 300 targets patients, focuses on adolescent patients who visit Babil Educational Hospital for Maternity and Children/Heredity Blood Disease Center on a regular basis. (Hereditary Blood

Disease Center, 2022).

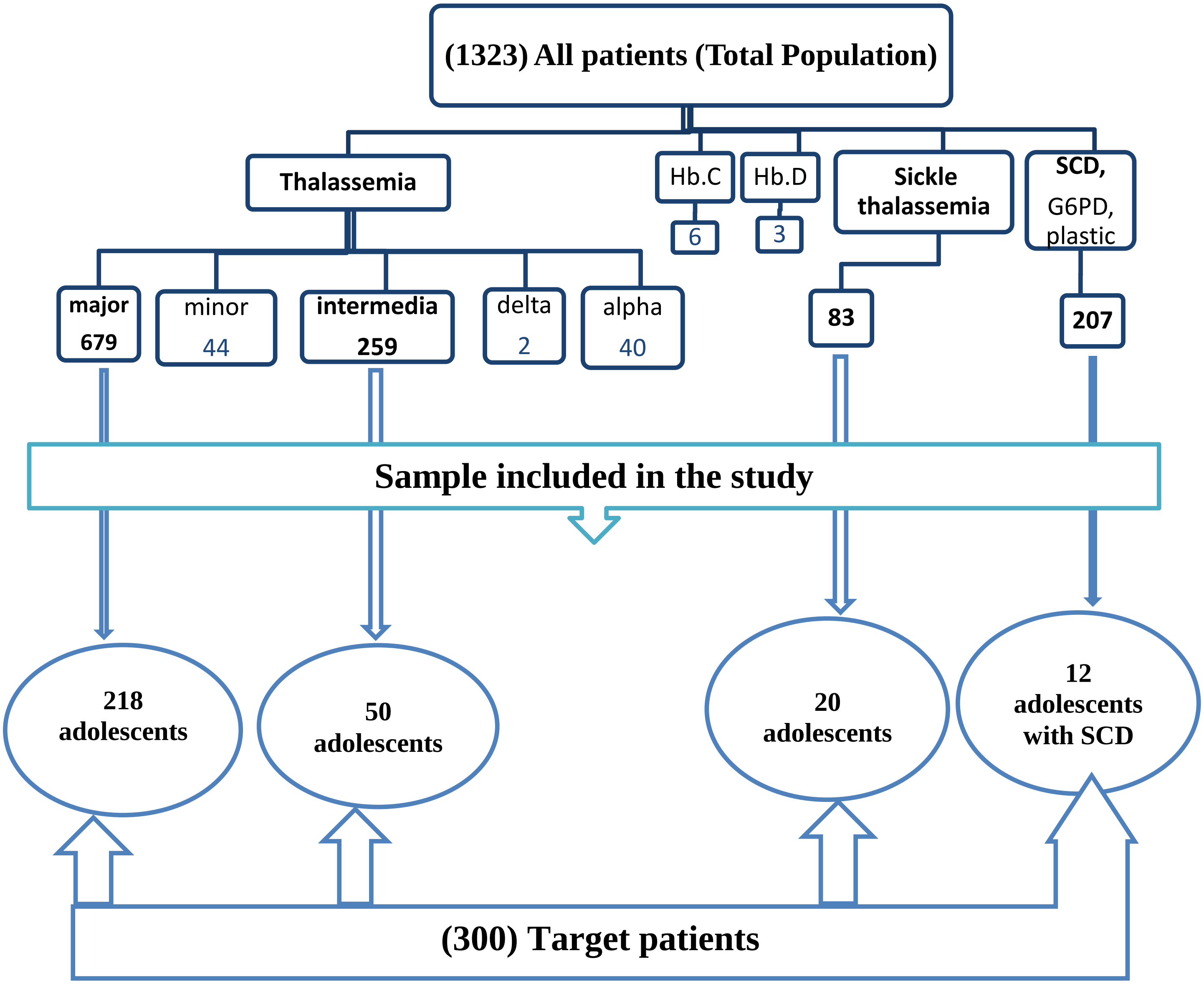

Figure (3-1) Distribution of the patients as a total population in the heredity blood disease center

3.4.1. Sample's Inclusion Criteria

Participants in the current study were required to meet certain requirements, include the following:

1. Adolescents, both boys and females, between the ages of 10 _ 19 years.
2. A resident of Babylon Province.

3. Patients diagnosed with Beta thalassemia major, intermediate thalassemia, sickle thalassemia, and sickle cell anemia.

3.4.2. Exclusion Criteria

1- Patients who are under 10 years and over 19 years old

2- Patients who refuse to take part in the research

3- Patients are diagnosed with other genetic blood diseases such as alpha thalassemia, delta thalassemia, aplastic anemia, and G6PD.

3.5. The Instrument of the Study

A questionnaire is modified for data collection to accomplish the goals of the current study after the comprehensive review of the relevant literature. It consist of five sections was designed to cover all aspects of the study as shown in Appendix (B).

Part I: Socio-Demographic Characteristics

This section is shown on a socio demographic data sheet consists of numerous items classified as adolescent's and their family’s information include: patient’s age, gender, order among siblings, and level of education (categorized into 4 levels; unable to read and write, primary, intermediate and secondary level). Parents education (categorized into 4 level also; unable to read and write, primary, secondary and diploma or above), occupation (categorized into 2 types employed and unemployed), parent consanguinity, economic status of family, and residency.

Part II: Medical History

This part contain multiple items about disease information such as type of illness (classified into 4 types of health disorders; major thalassemia, intermediate thalassemia, sickle beta thalassemia and sickle cell anemia), frequency of blood transfusion/ month, hemoglobin level, age at diagnosis, disease complication (such as osteoporosis, splenomegaly, hepatomegaly, hepatitis and diabetes), height and weight measurements by using electronic scale and tape measures, and sibling status of disease. The anthropometric measures of adolescents include (weight, height, and age). The key indications used to measure a child's growth over time are head circumference, weight, and measurements of length or height. Children under the age of two are normally measured by length, whereas those older than three are typically measured by height. The Centers for Disease Control and Prevention (CDC) and the World Health Organization (WHO) both recommend using growth charts regularly for evaluating growth. The charts in Appendix (D) are used for children and adolescents from 2-20 years of age (Rogol & Hayden, 2014).

Part III: Physical Wellness for Hemoglobinopathic Adolescents

This section includes 23 items about physical wellness classified as following manner: 3 items asked about physical health related to clothes, 5 items for physical health related to transferring, 4 items about physical health related to school, 3 items for physical health related to play, 6 items about physical health related to hygiene, and 2 item for physical health related to sleep.

Part IV: Psychological Wellness for Hemoglobinopathy Adolescents

This part contains organized elements concerning psychological wellness. It consists of 32 items include 7 items ask about somatic symptoms, 7 items for anxiety & insomnia, 7 items about social dysfunction, 7 items for severe depression, and 4 items for psychological or cognitive health related to school.

Part V: Social Wellness for Hemoglobinopathy Adolescents

Social aspects consisting of 12 items include relations of adolescent with family and friends, and adaptation with community.

3.6. Rating and Scoring

An instrument was performed using (3) levels form likert scale for measuring hemoglobinopathic adolescents’ physical and psychosocial wellness.

The instrument's score and rating 1= always, 2= sometimes, and 3= never.

3.7. Validity of the Study Instrument

It is achieved by a panel of 19 experts to examine the, relevance, adequacy and clarity of the questionnaire. A preparatory copy of the questionnaire is drawn up and submitted to nineteen experts with more than ten years' experience. They are (8) "faculty members from the college of nursing university of Babylon, (2) pediatricians and faculty members from the college of medicine /university of Babylon", (4) "from the college of nursing/ university of Baghdad, (2) faculty member from the college of nursing university of Kufa, (2) faculty member from the college of nursing/university of Karbala, and" lastly, (1) from faculty of nursing/the university of Thi-Qar. Suggestions and comments by the expertise were taken into account, as presented in appendix (C).

3.8. Pilot Study

The tool's first experiment was conducted on a group of (15) hemoglobinopathic adolescents chosen from the center specified for the current research, an interviewing technique application. During the period from 2nd February to 8th, 2022, the pilot research sample is excluded from the study's original sample.

It's done for a variety of reasons, including:

1. Evaluate the instrument's reliability.
2. Determine the nature of the difficulties that the researcher may face during the study.
3. Ensure that the instrument and its content are appropriate for the situation and identify the required modifications.
4. Estimated time needed for each patient for the interview and the measurements of (weight, height).

The findings demonstrated that:

- The questionnaire items were understood and clear as well.
- Minor changes are made on a few items.
- The time required to complete the instrument for each participant between (20-30 min).

3.9. Reliability of the Study Instrument

The data are gathered from 15 hemoglobinopathic patients to assess the internal instrument reliability. Internal consistency, reliability was computed via the calculation the cronbach's alpha technique which was = 0.879 as an accepted result in separated and total items. Reliability involves a research instrument's consistency in measuring a variable of interest. The questionnaire's determination of reliability is set up on the technique of "internal consistency / Alpha Cronbach", as calculated electronically by using (SPSS) version 26 of reliability analysis and shown in table below (3-1).

Table 3.1 Reliability of the Studied Questionnaire

Domains	No. of Items	Alpha Cronbach	Acceptable Value	Result
Physical Wellness	23	0.909	> 0.60	Accepted
Psychological Wellness	32	0.864	> 0.60	Accepted
Social Wellness	12	0.645	> 0.60	Accepted
Total	67	0.879	> 0.60	Accepted

3.10. Collection of the data

The data was collected through the period from 15th February to 15th April 2022; by utilizing the questionnaire and the interview technique with the participants who were interviewed singly at blood disease center, in Babylon province, using the Arabic version and by interviewing all participants inclusive in the research sample in a similar manner, the same questionnaire was used at the same place to achieve objectives of the research and other part collected by using tape measure for height and electronic scale for weight, which takes in estimation (20-30) minute for each participant.

3.11. Methods of Analysis

"Data analysis is done by using the statistical package for social sciences version (26)". The following methods of analysis were used, and the test results were measured and analyzed.

3.11.1. Descriptive Statistic

- (F) Frequencies, Percentages (%)
- Mean (x)
- Stander deviation (SD)
- The determining grades of the responses, subjects physical and psychosocial wellness with hemoglobinopathic disorders are divided into three grades (low, fair and high).

$$\textbf{Range of Score} = \frac{\textbf{Max}-\textbf{Min}}{\textbf{Rating}} = \frac{\textbf{3}-\textbf{1}}{\textbf{3}} = \textbf{0.66}$$

- 1 _ 1.66 = poor.
- 1.67 _ 2.33 = Fair.
- 2.34 _ 3 = well.

3.11.2. Inferential Analysis

a- The Cronbach Alpha Technique is form questionnaire reliability.

b- "Chi-square test": is used to determine the correlation between physical and psychosocial wellness of sample with demographic characteristics.

3.12. Obstacles of the Study

1. The researcher was unable to increase the number of the sample due to poor electronic documentation

2. As well as the lack of patient's follow-up at the center due to poor of their socioeconomic status, and far location of the center to their residency.

3. Weakness in the follow-up of patients by the health team and the disclosure of the reasons for their discontinuation from visiting those specialized government centers.

4. Some adolescents refused the participation in the present study.

CHAPTER FOUR
RESULT OF STUDY

CHAPTER FOUR

RESULT OF STUDY

This chapter presents the results of data analysis were organized in tables aligned to the study's objectives as follows:

Table 4.1:Distribution the demographic data of the sample(N =150)

Demographical data		Frequency	Percentage %
Age	10-13	59	**39.3**
	14-16	49	32.7
	17-19	42	28.0
	Total	150	100.0
	Mean (SD)	14.7 (± 2.82)	
Gender	Male	75	50.0
	Female	75	50.0
	Total	150	100.0
Patient order among siblings	1-3	96	**64.0**
	4-6	48	32.0
	7-9	6	4.0
	Total	150	100.0
Patient Level of education	Unable to read and write	11	7.3
	Primary level	65	**43.3**
	intermediate level	47	31.3
	Secondary level	27	18.0
	Total	150	100.0
Mother Education	Unable to read and write	17	11.3
	Primary level	88	**58.7**
	Secondary level	30	20.0
	Diploma or above	15	10.0
	Total	150	100.0

Father Education	Unable to read and write	14	9.3
	Primary level	73	**48.7**
	Secondary level	35	23.3
	Diploma or above	28	18.7
	Total	150	100.0
Mothers Occupation	Employed	19	12.7
	Unemployed	131	**87.3**
	Total	150	100.0
Fathers Occupation	Employed	131	**87.3**
	Unemployed	19	12.7
	Total	150	100.0
Parent Consanguinity	Yes	114	**76.0**
	No	36	24.0
	Total	150	100.0
Economic State of Family	Enough	51	34.0
	Enough to some extent	71	**47.3**
	Not enough	28	18.7
	Total	150	100.0
Residency	Urban	69	46.0
	Rural	81	**54.0**
	Total	150	100.0

Table 4.1: Shows that 59 (39.3%) of sample age were between (10-13) years, distributed between 75 (50 %) were male and 75 (50 %) were female with 96 (64%) were (1-3) patients order among their siblings and revealed as 65 (43.3%) were primary level of education. In relation to mother education, it represent 88 (58.7%) of them have primary level of education, as well as fathers from primary also as estimated with 73 (48.7%). Concerning to mothers occupation, 131 (87.3%) were Unemployed and 131 (87.3%) of

fathers occupation were employed.

This table above also publicized that 114(76%) were respond yes to Parent consanguinity. Regarding to economic state of family, less than half 71(47.3%) were Enough to some extent and more than half 81 (54%) from rural .

Table 4.2: Distribution of the sample according to their Medical Histories (N =150)

Medical History		Frequency	Percent
Type of illness	Major Thalassemia	121	**80.7**
	Intermediate thalassemia	19	12.7
	Sickle Beta Thalassemia	7	4.7
	Sickle cell anemia	3	2.0
	Total	150	100.0
Frequency of blood transfusion/ month	One time	66	**44.0**
	Twice	62	41.3
	Three times & over	22	14.7
	Total	150	100.0
Age at diagnosis	1-3	128	**85.3**
	4-5	12	8.0
	6-12	10	6.7
	Total	150	100.0
Osteoporosis	Yes	50	33.3
	No	100	**66.7**
	Total	150	100.0
Splenomegaly	Yes	80	**53.3**
	No	70	46.7
	Total	150	100.0
Hepatomegaly	Yes	30	20.0
	No	120	**80.0**
	Total	150	100.0
Hepatitis	Yes	10	6.7
	No	140	**93.3**
	Total	150	100.0

Diabetes	Yes	8	5.4
	No	142	**94.6**
	Total	150	100.0
Cardiomegaly	Yes	12	8
	No	138	**92.0**
	Total	150	100.0
Is there more than one affected child in the family	Yes	98	**65.3**
	No	52	34.7
	Total	150	100.0
BMI Percentile	Less than 5th percentile	33	22.0
	5th to less than 85th percentile	111	**74.0**
	85th to less than 95th percentile	5	3.3
	95th and more	1	.7
	Total	150	100.0

Table 4.2: Shows that 121 (80.7%) of children were major thalassemia and 66 (44 %) were take one frequency of blood transfusion/ month. Regarding to age at diagnosis 128 (85.3%) of the respondents were (1-3) years.

In relation to history of disease, about 100 (66.7%) were didn't have osteoporosis, while 80 (53.3%) having splenomegaly, 120 (80%) didn't have hepatomegaly, 140 (93.3%) didn't have hepatitis, 142 (94.6%) didn't have diabetes, and 138 (92%) didn't have cardiomegaly.

Concerning to BMI Percentile, approximately 111 (74 %) were between 5th to less than 85th percentile, and 98 (65.3%) of them having more than one affected child in the family.

Table 4.3:Distribution of sample's Hemoglobin level (N =150)

Mean	7.76
Std. Deviation	.874
Minimum	5.8
Maximum	10.5

Table 4.3 show that the minimum Hb were 5.8 while the Maximum were 10.5, the mean of Hb were 7.76 and standard deviation were 0.874.

Table 4.4: Sample's responses to Physical Wellness for Hemoglobinopathy Adolescents (N =150)

Items		F	%	M	SD	Ass.
1. Have difficulties in wearing my clothes	Always	7	4.7	2.74	.53	Well
	Sometime	25	16.7			
	Never	118	78.7			
	Total	150	100.0			
2. Need help from other to keep and arrange my clothes	Always	75	50.0	1.82	.89	Fair
	Sometime	26	17.3			
	Never	49	32.7			
	Total	150	100.0			
3. Have difficulties to care of my general appearance	Always	23	15.3	2.40	.74	Well
	Sometime	43	28.7			
	Never	84	56.0			
	Total	150	100.0			
4. Have difficulty in walking alone	Always	13	8.7	2.33	.63	Fair
	Sometime	74	49.3			
	Never	63	42.0			
	Total	150	100.0			
5. Lift the heavy things with difficulty.	Always	107	71.3	1.38	.65	Poor
	Sometime	29	19.3			
	Never	14	9.3			
	Total	150	100.0			
6. Have efforts in going to school	Always	54	36.0	1.89	.77	Fair
	Sometime	58	38.7			
	Never	38	25.3			
	Total	150	100.0			
7. Have fatigue and generalized weakness	Always	25	16.7	1.93	.51	Fair
	Sometime	110	73.3			
	Never	15	10.0			
	Total	150	100.0			
8. Need help from other to perform the sport activities	Always	57	38.0	1.92	.82	Fair
	Sometime	48	32.0			
	Never	45	30.0			

	Total	150	100.0			
9. Have school absenteeism because of tired	Always	62	41.3	1.8	.76	Fair
	Sometime	56	37.3			
	Never	32	21.3			
	Total	150	100.0			
10. Perform my school homework with difficulty due to fatigue	Always	53	35.3	1.84	.72	Fair
	Sometime	68	45.3			
	Never	29	19.3			
	Total	150	100.0			
11. My attention drops during the lesson due to fatigue	Always	53	35.3	1.86	.74	Fair
	Sometime	64	42.7			
	Never	33	22.0			
	Total	150	100.0			
12. Have exertion to go to the school-picnics	Always	70	46.7	1.84	.87	Fair
	Sometime	33	22.0			
	Never	47	31.3			
	Total	150	100.0			
13. Have efforts in playing with other children	Always	35	23.3	2.19	.79	Fair
	Sometime	51	34.0			
	Never	64	42.7			
	Total	150	100.0			
14. Find it hard to do my favorite hobbies	Always	20	13.3	2.46	.72	Well
	Sometime	41	27.3			
	Never	89	59.3			
	Total	150	100.0			
15. I'm looking for other activities that don't require a lot of energy	Always	81	54.0	1.59	.71	Poor
	Sometime	49	32.7			
	Never	20	13.3			
	Total	150	100.0			
16. Have difficulties in taking bath or shower by my self	Always	8	5.3	2.71	.55	Well
	Sometime	27	18.0			
	Never	115	76.7			
	Total	150	100.0			
17. Find struggle in drying my	Always	3	2.0	2.84	.41	Well
	Sometime	18	12.0			

body	Never	129	86.0			
	Total	150	100.0			
18. Need help from others to prepare my hair style	Always	29	19.3	2.4	.79	Well
	Sometime	31	20.7			
	Never	90	60.0			
	Total	150	100.0			
19. Require help by others in toilet training	Sometime	6	4.0	**2.96**	.19	Well
	Never	144	96.0			
	Total	150	100.0			
20. Need help by others in fingernails care	Always	14	9.3	2.72	.62	Well
	Sometime	14	9.3			
	Never	122	81.3			
	Total	150	100.0			
21. I have trouble in brushing teeth	Always	8	5.3	2.70	.56	Well
	Sometime	28	18.7			
	Never	114	76.0			
	Total	150	100.0			
22. Go to bed regularly	Never	53	35.3	2.01	.85	Well
	Sometime	42	28.0			
	Always	55	36.7			
	Total	150	100.0			
23. Have distress in sleeping	Always	27	18.0	2.21	.72	Fair
	Sometime	64	42.7			
	Never	59	39.3			
	Total	150	100.0			

Cut of points value(0.66): Poor = (1-1.66), Moderate = (1.67-2.33), Good = (2.34-3).

M= mean, SD=standard deviation, Freq=frequency, % = percentage.

Table 4.4: Shows that the maximum mean was 2.96 with the item (Require help by others in toilet training) while a minimum mean 1.38 with the item (Lift the heavy things with difficulty).

Table 4.5:Overall level of Physical Wellness among Hemoglobinopathy Adolescents (N =150).

Physical Wellness	Frequency	Percentage%	Mean	SD
Poor	18	12.0	50.58	7.6
Fair	86	**57.3**		
Well	46	30.7		
Total	150	100.0		

Poor = (23-38), Moderate = (39-54), Good = (55-69), SD=standard deviation

Table 4.5 show that 86 (57.3%) of sample having moderate physical wellness for hemoglobinopathy adolescents, and 46(30.7 %)of sample having good physical wellness , whereas, 18(12.0%) of them having poor level for physical wellness.

Table 4.6:Distribution of Responses of Psychological Wellness for Hemoglobinopathy Adolescent (N =150)

Items		Frequency	Percent	M	SD	Ass.
1. Suppose perfect well even in good health	Never	16	10.7	2.07	.53	Fair
	Sometime	107	71.3			
	Always	27	18.0			
	Total	150	100.0			
2. Think in need for good tonic	Always	44	29.3	2.12	.83	Fair
	Sometime	43	28.7			
	Never	63	42.0			
	Total	150	100.0			
3. Feel tired and exhausted	Always	22	14.7	1.92	.46	Fair
	Sometime	117	78.0			
	Never	11	7.3			
	Total	150	100.0			
4. Feel sick	Always	21	14.0	2.0	.53	Fair
	Sometime	108	72.0			
	Never	21	14.0			
	Total	150	100.0			
	Always	28	18.7	1.96	.58	Fair

5. Suffer from a headache	Sometime	99	66.0			
	Never	23	15.3			
	Total	150	100.0			
6. Feel tightness and pressure in my head	Always	8	5.3	2.59	.59	Fair
	Sometime	45	30.0			
	Never	97	64.7			
	Total	150	100.0			
7. Feel sudden states of cold or heat	Always	23	15.3	2.1	.63	Fair
	Sometime	89	59.3			
	Never	38	25.3			
	Total	150	100.0			
8. I have decrease in my sleeping hours due to anxiety	Always	15	10.0	2.46	.67	Well
	Sometime	51	34.0			
	Never	84	56.0			
	Total	150	100.0			
9. If I fall asleep, I find it hard to stay asleep	Always	30	20.0	2.28	.77	Fair
	Sometime	48	32.0			
	Never	72	48.0			
	Total	150	100.0			
10. Feel tired all the time	Always	17	11.3	2.28	.65	Fair
	Sometime	74	49.3			
	Never	59	39.3			
	Total	150	100.0			
11. Get nervous quickly and in a bad mood	Always	73	48.7	1.66	.72	Poor
	Sometime	55	36.7			
	Never	22	14.7			
	Total	150	100.0			
12. Feel scared or panicky for no good reason	Always	34	22.7	2.3	.81	Fair
	Sometime	36	24.0			
	Never	80	53.3			
	Total	150	100.0			
13. Feel like I carry the worries of the world on My head	Always	23	15.3	2.37	.73	Well
	Sometime	48	32.0			
	Never	79	52.7			
	Total	150	100.0			
14. Feel nervous and	Always	39	26.0	2.04	.75	Fair
	Sometime	65	43.3			

strung-up all the time	Never	46	30.7			
	Total	150	100.0			
15. Feel like I can finish myself with certain things	Never	22	14.7	2.32	.71	Fair
	Sometime	57	38.0			
	Always	71	47.3			
	Total	150	100.0			
16. Taking longer over the things I do	Always	50	33.3	2.04	.84	Fair
	Sometime	44	29.3			
	Never	56	37.3			
	Total	150	100.0			
17. Feel on the whole I'm doing things well	Never	13	8.7	2.52	.65	Well
	Sometime	45	30.0			
	Always	92	61.3			
	Total	150	100.0			
18. I am satisfied with the way I've carried out my task	Never	11	7.3	2.53	.63	Well
	Sometime	48	32.0			
	Always	91	60.7			
	Total	150	100.0			
19. Feel playing a useful part in things	Never	19	12.7	2.4	.704	Well
	Sometime	52	34.7			
	Always	79	52.7			
	Total	150	100.0			
20. Can make decisions	Never	42	28.0	2.1	.81	Fair
	Sometime	50	33.3			
	Always	58	38.7			
	Total	150	100.0			
21. I am able to enjoy my normal day-to-day activities	Never	21	14.0	2.37	.71	Well
	Sometime	52	34.7			
	Always	77	51.3			
	Total	150	100.0			
22. Feel like a worthless person	Always	10	6.7	2.64	.605	Well
	Sometime	34	22.7			
	Never	106	70.7			
	Total	150	100.0			
23. Felt that life is entirely hopeless	Always	19	12.7	2.54	.71	Well
	Sometime	30	20.0			
	Never	101	67.3			
	Total	150	100.0			

24. Feel that life is not worth living	Always	20	13.3	2.54	.719	Well
	Sometime	28	18.7			
	Never	102	68.0			
	Total	150	100.0			
25. Have the idea of excluding my life	Always	6	4.0	2.82	.47	Well
	Sometime	15	10.0			
	Never	129	86.0			
	Total	150	100.0			
26. Found at times I couldn't do anything because my nerves were too bad	Always	43	28.7	2.01	.76	Fair
	Sometime	63	42.0			
	Never	44	29.3			
	Total	150	100.0			
27. Wish I was dead and away from everything around me	Always	25	16.7	2.55	.76	Well
	Sometime	17	11.3			
	Never	108	72.0			
	Total	150	100.0			
28. Find the idea of getting rid of my life on my mind a lot	Always	6	4.0	2.80	.49	Well
	Sometime	18	12.0			
	Never	126	84.0			
	Total	150	100.0			
29. Have trouble in understanding my lessons	Always	54	36.0	1.83	.72	Fair
	Sometime	67	44.7			
	Never	29	19.3			
	Total	150	100.0			
30. I am distressed in coping in school and other Pupils	Always	42	28.0	2.16	.83	Fair
	Sometime	42	28.0			
	Never	66	44.0			
	Total	150	100.0			
31. Have difficulties in understanding my school administration about my health status	Always	47	31.3	2.16	.87	Fair
	Sometime	32	21.3			
	Never	71	47.3			
	Total	150	100.0			
32. Achieve low grades in school-examinations	Always	49	32.7	1.87	.716	Fair
	Sometime	71	47.3			
	Never	30	20.0			
	Total	150	100.0			

Cut of points value(0.66): Poor = (1-1.66), Moderate = (1.67-2.33), Good = (2.34-3).

M= mean, SD=standard deviation, Freq=frequency, % = percentage.

Table 4.6: Shows that the maximum mean was 2.82 with the item (Have the idea of excluding my life) while a minimum mean 1.66 with the

item (Get nervous quickly and in a bad mood).

Table 4.7: Overall level of Psychological Wellness among Hemoglobinopathy Adolescents (N =150)

Psychological Wellness	Frequency	Percent	Mean	SD
Poor	6	4.0	72.44	9.64
Fair	59	39.3		
Well	85	**56.7**		
Total	150	100.0		

Poor = (32-53), Fair = (54-75), Well = (76-96), SD=standard deviation

Table 4.7 show that 85 (56.7%) of sample having a good Psychological wellness for hemoglobinopathy adolescent. While (39.3%) of them having moderate responses, and (4.0%) having poor responses.

Table 4.8:Distribution responses of social Wellness among Hemoglobinopathy Adolescents (N =150)

Items		Frequency	Percent	M	SD	Ass.
1. Get emotional help and support I need from my family	Never	5	3.3	2.66	.54	Well
	Sometime	41	27.3			
	Always	104	69.3			
	Total	150	100.0			
2. My family is willing to help me make decisions	Never	4	2.7	**2.70**	.51	Well
	Sometime	36	24.0			
	Always	110	73.3			
	Total	150	100.0			
3. Can talk about my problem with my family	Never	52	34.7	1.99	.83	Fair
	Sometime	47	31.3			
	Always	51	34.0			
	Total	150	100.0			
4. My family support me to participate in social activities	Never	31	20.7	2.31	.79	Fair
	Sometime	41	27.3			
	Always	78	52.0			
	Total	150	100.0			
5. Get the same attention from my father and mother	Never	11	7.3	2.68	.603	Well
	Sometime	25	16.7			
	Always	114	76.0			
	Total	150	100.0			

6. There is special person in my life who care about my feelings	Never	30	20.0	2.40	.802	Well
	Sometime	30	20.0			
	Always	90	60.0			
	Total	150	100.0			
7. Find it difficult to make friends	Always	40	26.7	2.21	.84	Fair
	Sometime	38	25.3			
	Never	72	48.0			
	Total	150	100.0			
8. Find myself isolated among people	Always	39	26.0	2.23	.83	Fair
	Sometime	37	24.7			
	Never	74	49.3			
	Total	150	100.0			
9. My illness affects my relationship with my family and friends	Always	25	16.7	2.47	.765	Well
	Sometime	29	19.3			
	Never	96	64.0			
	Total	150	100.0			
10. Suffer from a lack of adaptation to disease and society	Always	64	42.7	1.74	.72	Fair
	Sometime	61	40.7			
	Never	25	16.7			
	Total	150	100.0			
11. Find it difficult to speak in front of others	Always	39	26.0	2.10	.78	Fair
	Sometime	57	38.0			
	Never	54	36.0			
	Total	150	100.0			
12. My parents don't listen to me when I talk	Always	16	10.7	2.62	.67	Well
	Sometime	25	16.7			
	Never	109	72.7			
	Total	150	100.0			

Cut of points value(0.66): Poor = (1-1.66), Moderate = (1.67-2.33), Good = (2.34-3).

M= mean, SD=standard deviation, Freq=frequency, % = percentage.

Table 4.8: Shows that the maximum mean was 2.70 with the item (My family is willing to help me make decisions) while a minimum mean 1.74 with the item (Suffer from a lack of adaptation to disease and society).

Table 4.9: Overall level of social Wellness among Hemoglobinopathy Adolescents (N =150).

Social Wellness	Frequency	Percent	Mean	SD
Poor	22	14.7	28.14	4.31
Fair	42	28.0		
Well	86	**57.3**		
Total	150	100.0		

Poor = (12-20), Fair = (21-28), Well = (29-36), SD=standard deviation

Table 4.9 show that 86 (57.3%) of sample having a good social wellness for hemoglobinopathy adolescent. While (28.0%) having moderate responses, and (14.7%) having poor responses for social wellness.

Table 4.10: Correlation between Psychological Wellness and Physical Wellness.

		Psychological Wellness			**Total**	**Correlation**	
		Poor	Moderate	Good		r	Sig
Physical Wellness	Poor	4	11	3	18	0.487^{**}	**0.001**
	Moderate	2	42	42	86		
	Good	0	6	40	46		
Total		6	59	85	150		

**Correlation is significant at the 0.001 level.
*Correlation is significant at the 0.05 level.
r= pearson correlation
P-value ≤ 0.05

Table 4.10 demonstrated that strong significant correlation between physical wellness and psychological wellness at P-value ≤ 0.05.

Table 4.11: Correlation between Social Wellness and Physical Wellness.

		Social Wellness			**Total**	**Correlation**	
		Poor	Moderate	Good		r	Sig
Physical Wellness	Poor	8	5	5	18	0.277^{*}	**0.002**
	Moderate	10	27	49	86		
	Good	4	10	32	46		
Total		22	42	86	150		

**Correlation is significant at the 0.001 level.
*Correlation is significant at the 0.05 level.
r= pearson correlation
P-value ≤ 0.05

Table 4.11, showing a significant correlation between physical wellness and social wellness at P-value ≤ 0.05.

Table 4.12: The correlation between social wellness and psychological wellness

		Social Wellness			**Total**	**Correlation**	
		Poor	Moderate	Good		r	Sig
Psychological Wellness	Poor	5	1	0	6	0322^{**}	**0.001**
	Moderate	9	21	29	59		
	Good	8	20	57	85		
Total		22	42	86	150		

**Correlation is significant at the 0.001 level.

*Correlation is significant at the 0.05 level.

r= pearson correlation

P-value ≤ 0.05

Table 4.12 reveals a significant correlation between psychological wellness and social wellness "at P-value ≤ 0.05".

Table 4.13: The associations between Physical Wellness and their demographic data

Demographical data	X^2	DF	p-value
Age	18.003^{**}	4	**.001**
Gender	4.061	2	.131
patient Level of education	45.149^{**}	6	**.001**
patient order among siblings	4.660	4	.324
Mother Education	32.24^{**}	6	**.001**
Father Education	13.883^{*}	6	**.031**
Mothers occupation	2.493	2	.288
Fathers occupation	.396	2	.820
Parent consanguinity	7.611^{*}	2	**.022**
Economic state of Family	8.300	4	.081
Residents	3.266	2	.195
X^2 = Chi-square	** significant at the 0.001 level.		
P value ≤ 0.05	* significant at the 0.05 level.		

Table 4.13 demonstrate there is a high significant association between physical Wellness and several demographical data at a ($p \leq 0.05$), like (age, patient level of education, mother education, father education, and parent consanguinity). Except with (gender, patient order among siblings, mothers' occupation, fathers' occupation, economic state of family, and residents) there is a non-significant association with physical Wellness at P-value ≤ 0.05.

Table 4.14: The association between Psychological Wellness and childs' demographic data.

Demographical data	X^2	DF	p-value
Age	1.088	4	.896
Gender	4.706	2	.095
patient Level of education	14.379^{*}	6	**.026**
patient order among siblings	3.771	4	.438
Mother Education	8.731	6	.189
Father Education	7.965	6	.241

Mothers occupation	1.066	2	.587
Fathers occupation	3.501	2	.174
Parent consanguinity	2.293	2	.318
Economic state of Family	12.856*	4	**.012**
Residents	3.280	2	.194

X2 = Chi-square ** significant at the 0.001 level.
P value ≤ 0.05 * significant at the 0.05 level.

Table 4.14 reveals a highly significant association between psychological wellness and several demographical data at P. ≤ (0.05), like (patient level of education, and the economical state of the family). Except with (age, gender, patient order among siblings, Mothers occupation, Fathers occupation, Mother Education, Father Education, Parent consanguinity, Residents) there is a non-significant association with psychological wellness at a P-value ≤ 0.05.

Table 4.15: The association between social wellness and childs' demographical data.

Demographical data	**X^2**	**DF**	**p-value**
Age	7.509	4	.111
Gender	.913	2	.633
patient Level of education	17.124**	6	**.009**
patient order among siblings	3.803	4	.433
Mother Education	18.866**	6	**.004**
Father Education	10.668	6	.099
Mothers occupation	2.701	2	.259
Fathers occupation	2.992	2	.224
Parent consanguinity	1.292	2	.524
Economic state of Family	14.794**	4	**.005**
Residents	1.511	2	.470

X2 = Chi-square ** high significant at the 0.001 level.
P value ≤ 0.05 * significant at the 0.05 level.

Table 4.15 reveals a high significant association between social wellness and several demographical data at p ≤ (0.05), like (patient level of education, mothers occupation, and the economical state of the family). except with (age, gender, patient order among siblings, fathers occupation, mother education, father education, parent consanguinity, and residents) there is a non-significant association with social wellness at P-value ≤ 0.05.

Table 4.16: The association between physical wellness and their medical history

Medical History	**X^2**	**DF**	**p-value**
Type of illness	9.251	6	.160
Frequency of blood transfusion/ month	3.907	4	.419
Age at diagnosis	.916	4	.922
Osteoporosis	.267	2	.875
Splenomegaly	1.585	2	.453
Hepatomegaly	3.438	2	.179
Hepatitis	2.616	4	.624
Diabetes	6.366	4	.173
Cardiomegaly	6.359	4	.174
Is there more than one affected child in the family	.229	2	.892
BMI percentile	2.251	6	.895
X^2 = Chi-square P value ≤ 0.05	** high significant at the 0.001 level. * significant at the 0.05 level.		

Table 4.16 demonstrate there is a non-significant association between Physical Wellness and any medical history at a P-value ≤ (0.05).

Table 4.17: The association between psychological wellness and their history of disease

Medical History	**X^2**	**DF**	**p-value**
Type of illness	39.391**	6	**.001**
Frequency of blood transfusion/ month	7.769	4	.100
Age at diagnosis	1.727	4	.786
Osteoporosis	5.353	2	.069
Splenomegaly	7.576*	2	**.023**
Hepatomegaly	6.298*	2	**.043**
Hepatitis	2.834	4	.586
Diabetes	2.731	4	.604
Cardiomegaly	8.930	4	.063
Is there more than one affected child in the family	1.546	2	.462
BMI percentile	6.863	6	.334
X^2 = Chi-square P value ≤ 0.05	** significant at the 0.01 level. * significant at the 0.05 level.		

Table 4.17 show there are a significant and highly significant association between Psychological Wellness and some medical history at $p \leq 0.05$, such as (Type of illness, Splenomegaly, Hepatomegaly). Except with (Frequency of blood transfusion/ month, Age at diagnosis, Osteoporosis, Hepatitis, Diabetes, Cardiomegaly, Is there more than one affected child in the family, BMI percentile) that there is a non-significant association with psychological wellness at **P-value ≤ 0.05.**

Table 4.18: The association between social wellness and history of disease among the respondents.

History of disease	**X^2**	**DF**	**p-value**
Type of illness	10.824	6	.094
Frequency of blood transfusion/ month	3.807	4	.433
Age at diagnosis	2.861	4	.581
Osteoporosis	.204	2	.903
Splenomegaly	.343	2	.842

Hepatomegaly	1.133	2	.568
Hepatitis	2.956	4	.565
Diabetes	.751	4	.945
Cardiomegaly	2.553	4	.635
Is there more than one affected child in the family	3.646	2	.162
BMI percentile	8.403	6	.210

X^2 = Chi-square ** high significant at the 0.001 level.
P value ≤ 0.05 * significant at the 0.05 level.

Table 4.18 reveals a high significant association between social wellness and some medical history at *p* ≤ 0.05, such as (Type of illness, Splenomegaly, Hepatomegaly , with exception to (Frequency of blood transfusion/ month, Age at diagnosis, Osteoporosis, Hepatitis, Diabetes, Cardiomegaly, Is there more than one affected child in the family, BMI percentile) which indicate a non-significant association with social wellness at “***P-value ≤ 0.05”.***

Table 4.19: The correlations between hemoglobin and (physical psychological, and social wellness).

Variables	Hemoglobin	
	r	Sig
Physical Wellness	.191*	**0.027**
Psychological Wellness	.025	0.761
Social Wellness	.071	0.386

**Correlation is significant at the 0.01 level.
*Correlation is significant at the 0.05 level.
r= person correlation
P-value ≤ 0.05

Table 4.19 illustrate there is a significant correlation between Hemoglobin and the three variables(physical , psychological and social) whereas a highly significance effected by hemoglobinopathies at ***P-value ≤ 0.05***.

CHAPTER FIVE

DISCUSSION

CHAPTER FIVE

DISCUSSION

This chapter presents the discussion and interpretation of the study results, which targeted in a systematic way toward the study's objectives and supported by certain articles.

5.1. Discussion of the Sociodemographic Characteristic of the Adolescents and Their Families.

Throughout the data analysis, the study shows that the nature of the study sample is described by demographic characteristics of hemoglobinopathic adolescents.

Table 4-1 indicates that most of the adolescents age group are within 10_13 years old with a mean age is (14.7), and equally divided between males and females. This result go in line with the finding of (Issa *et al*.,2018) that carried out on 100 thalassemic adolescents aged 10_18 years, the demographic characteristics of this study show a majority age group is 10_14 years with high percentage prevalence. In a mixed-methods study design done by (Kambasu *et al*.,2019), among 140 patients with SCD their age was 8_17 years in order to assess (Health related quality of life of adolescents with SCD in sub-Saharan Africa: a cross-sectional study) which found that mean age was (14.25) years.

In case-control study a group of fifty patient with thalassemia major and fifty normal children within age 5_17 years were included by (Elzaree *et al*.,2018) about the Adaptation Functions and Psychosocial Issues among Children with Major Beta-Thalassemia had half of sample was male and other half was female.

Furthermore, supportive evidence is a descriptive study done by

(Shareef& Obaid,2017), on 50 patients in order to assess the knowledge of adolescents with thalassemia major regarding iron chelating therapy also found equally distributed of sample according to gender. In relation to the Patient order among siblings in the current study majority of them are between first and third child, which is supported by (Saeed &Aldoori,2020) that conducted in in the Babylon province, who showed that most of adolescent’s order in their families are between first and second child.

Regarding the patient level of education in the existing study most of them were primary level. Sustained by (Ishfaq *et al*.,2018), who carried out a cross sectional study on 200 major thalassemic patients in Pakistan as majority of them was primary level of education. Other study conducted by (Batool *et al*.,2017), who carried out a cross sectional study design on 91 patients with thalassemia major having age 12_18 years most of them was primary educational level.

In case-control survey was conducted in Minia University children’s hospital on 64 patients and 64 healthy children aged between 8_18 years were done by (Hakeem *et al*.,2018) about the Health-related quality of life among children and adolescent patients with transfusion-dependent ß-thalassemia in upper Egypt, had majority of patients (48.4%) were primary level of education.

Parents’ level of education in the present study more than half of them were primary level. These findings agree with result of (Saeed &Aldoori,2020), who carried out a cross sectional descriptive study design on 100 thalassemic and 100 healthy adolescents was carried to compare the psychosocial development of thalassemic and healthy adolescents in the Babylon province, had the uppermost of the study sample parents graduated from primary school. Another study done by (Baiee *et al*., 2015), also they found majority of the participants were parents who graduated from primary school.

Whereas, Quasi-experimental study conducted by (Hassan & Azzab, 2016) who studied (23) children for studying the impact of health instructions on the quality of life and psychological difficulties among thalassemic children. It shows that the highest percentage (43.5%) of mother's education were illiterate, while fathers education level (43.3%) was secondary or above.

Finding of the parent’s occupation in the current study shows majority for mothers are unemployed and fathers are employed. Supportive evidence is provided by (Saeed &Aldoori,2020), they found that majority mothers of thalassemic participants were unemployed, and majority of fathers were employed. This finding also agree with result of study that done with (Issa *et al.*,2018), they found the vast majority of mothers were housewife. This may be rationalized as due to the culture of the society from which the sample was collected does not accept the job for the women and do not allow them to complete their educations, especially if she has children, and in particular those who are sick and in need of continuous and chronic follow-up.

The present study also reveals that more than three quarters were respond yes to parent consanguinity. This finding agrees with result of study conducted by (Abdulsattar & Hattab,2017), is a descriptive study design was carried to assess the physical, psychosocial, and financial burdens of major beta thalassemia and its treatment upon (80) adolescents whose age range between 11_21 years old. At the thalassemia center in the Ibn albeldy pediatric and maternal teaching hospital. Had majority of participant have parent consanguinity. Another study included by (Al-Hakeim *et al.*,2020), on (1122 patients, aged 5 to 65 years old) who studied the “Hereditary Hematologic

Disorders in Najaf Province-Iraq'' found the consanguinity rate in their patients' parents (78.24%). From the perspective of the researchers marriage between relatives has been identified as a significant role in genetic illnesses, which are prevalent in the Iraqi populations.

Regarding to economic state of family, in the current study less than half were enough to some extent and residency of more than half from rural. This result supported by (Saeed &Aldoori,2020), they found economic state for most of the study sample were enough to some extent and the residence area of most thalassemic study sample was rural. Other study done by (Yengil *et al*., 2014) which conducted on (88) thalassemic patients and (63) caregivers of patients. It concluded that anxiety, depression, and QoL for sufferers and their caretakers', despite of the most of thalassemic patients from rural, whereas in another study held by (Aziz *et al*., 2013), which conducted on (140) mother's having thalassemic child which entitled, "Assessment of mother's practices toward their thalassemic children who receiving deseferal therapy" showed that most of thalassemic patients from urban areas.

5.2. Discussion of the medical history for the adolescent's patients

The present study in the table (4.2) shows that more than three-quarters of children had major thalassemia. This result agrees with the finding of a study conducted by (Abd *et al*., 2020) who studied (601) hemoglobinopathic patients titled (Hemoglobinopathies in Thi-Qar Governorate According to Blood Groups) and found that more than three-quarters of the sample had thalassemia. Another study done by (Al-Hakeim *et al*.,2020), also found thalassemia major is more prevalent than other types of hemoglobinopathic disorders.

From the researcher's point of view, these results could refer to the prevalence of thalassemia in Iraq being more than in other types of hemoglobinopathy disorders, this may be attributed to increasing the rate of consanguinity marriage, or neglect of the importance of marriage screening tests by people who are about to get married, especially those who live in the rural area and who have a lack of culture and healthy child development awareness as well as the effect of hemoglibinopathic disorders on their children personalities and body images.

Regarding to frequency of blood transfusion the current study shows more than two third of them were take one blood transfusion/ month. This result supported with the finding of cross-sectional case control study done by (Kumaravel *et al*.,2017), On 32 thalassemic children from 5 to 15 years and 32 healthy children matched with same age and sex, had transfusion frequency for majority of thalassemic children once monthly.

Another study conducted by (Venty *et al*.,2018), this cross-sectional study included (64) patients with thalassemia major aged 7 to <18 years. Which entitled a "Depression in children with thalassemia major: prevalence and contributing factors" who had a majority of participant takes one blood transfusion in months. Whereas, a cross-sectional study was conducted by (Mansoor *et al*.,2018) on (81) thalassemic adolescents, found more than half of sample was takes blood transfusion once every 2 weeks.

Regarding to age at diagnosis, in the present research vast majority of sample were (1-3) years. In case-control study done by (Mikael& Al-Allawi,2018) was conducted among 100 thalassemic children aged 6-18 years and 100 healthy from same age groups, in titled, (Factors impacting the quality of life for thalassemic children and adolescents in Iraqi Kurdistan) they found age of diagnosis for majority of patients from 1 to 3 years. Another study done by (Mansoor *et al*.,2018), they found the age of diagnosis for three-quarters of the sample was 0-12 month. Researchers opinion that the age of disease diagnosis has been early as possible, and they believe that may be early detection of disease help in controlling strenuous complications.

In relation to history of disease, disease complication in the current study about two third were didn’t have osteoporosis, more than half were having splenomegaly, more than three quarters were didn’t have hepatomegaly, and vast majority of them were didn’t have hepatitis, diabetes, and cardiomegaly. More supportive evidence is provided by (Abdulsattar & Hattab,2017), they depict in their study that patients experience many long-term complications such as hepatitis, growth retardation and splenomegaly. This rationalized to patients with thalassemia major maybe has a higher risk for recurrence of complications than other types of hemoglobinopathies

Regarding to more than one affected child in the family the present study shows that majority of hemoglobinopathic patients have sibling affected with disease. Supportive evidence is provided by (Saeed &Aldoori,2020), who conducted across sectional study on 100 thalassemic adolescents and 100 healthy with same age groups, they found in their study most thalassemic patient have sibling affected with disease. Furthermore, a study directed by (Kumaravel, et al., 2017), who carried out a cross sectional case control study on 32 thalassemic children and (32) controls matched to the age and sex. In the Thalassemia children's group majority of thalassemic patients have a sibling with thalassemia.

Concerning to BMI Percentile, the current research shows less than three quarters were between 5th to less than 85th percentile. This finding agrees with result of study done by (Saeed &Aldoori,2020), they found the majority of patients were 5th percentile while the majority of healthy adolescents are normal in weight, height, and BMI. The outcome of the contemporary study consistent with an Indian recent article, the conclusion of this article was that Hemoglobinopathies patients are short, have lower rates of growth and BMI, and their puberty spurts are either delayed or missing due to sub-optimal iron chelation therapy, low hemoglobin and high levels of ferritin (Saxena, 2017).

The researcher's point of view is that children with hemoglobinopathies have low growth than normal children in the same age group, this may be due to the negative effect of the disease on growth hormone for the affected children. It can be affected by the educational level of mothers who do not have awareness of the problems resulting from iron deposits, which affects the growth and development of children with genetic blood diseases.

However table 4.3 show that the minimum Hb was 5.8 while the Maximum was 10.5, and the mean of Hb was 7.76. This result borders in line with the finding of the study done by (Elzaree *et al*.,2018), included in a case-control study on a set of 50 children with thalassemia and 50 healthy children of the same age and sex, they find the mean of Hb was 7.79. Another study conducted by (Venty *et al*., 2018), on 64 thalassemic children aged 7_18 years, there finding the mean Hb level was 7.18.

5.3: Discussion of the responses of Physical wellness for hemoglobinopathic adolescents

The Study has revealed that all the domains of the physical wellness (physical health related to clothing, transferring, school activities, playing, personal hygiene, and sleeping) It had distribution on three scale levels (always, sometime, and never), according to (table 4.5) more than half of sample having moderate physical wellness for hemoglobinopathy adolescents.

Kshaish & Aziz, (2020), carried out a descriptive study about assessment of daily living activities for (100) thalassemic school age children in Baghdad city, they also find most of children are affected moderately in all domains of daily living activities.

A cross-sectional study directed by (Moyen *et al*., 2021), on 201 patients with SCD aged (6_19 years old) in order to determine the (Psychological Experience for Children with Homozygous SCD in Brazzaville) which found that the impact of disease on the patient's daily life was negative in all cases.

Regarding to physical health related to clothing, results of the current study shown that more than three quarters of sample never have difficulties in wearing clothes, half of them always need help from other to keep and arrange clothes, and more than half were never have difficulties to care of general appearance. In descriptive study done by (Kshaish & Aziz, 2020), they found (68%) of school age children sometimes wearing clothes difficultly, (54%) never needs to help from other to keep and arrange their clothes, and (70%) Sometimes have difficulties to care their general appearance.

Concerning to physical health related to transferring in the present study table (4.4) shown that less than half of sample sometime have difficulty in walking alone, more than thirds of them always lift the heavy things with difficulty, less than two fifth of them sometimes have efforts in going to school, more than two third sometimes have fatigue and generalized weakness, and less than two fifth always need help from other to perform the sport activities. This result supported with the findings of descriptive qualitative research approach conducted by (Akter *et al*.,2020), which recruit purposively 20 children with thalassemia, in order to assess (the living experience of children with thalassemia in Bangladesh) they found that majority of kids stated that they were having difficulty completing the task.

feeling tired when do any work, and can’t do heavy work. Felling weak in walking. they are feeling tired when they go for playing the games, and feeling tired when they go to school.

As well as the study done by (Kshaish & Aziz, 2020), they found (63%) Sometimes have difficulty in walking alone, (64%) sometimes lift the heavy things with difficulty, (58%) sometimes have difficulties in going to school, (50%) sometimes having fatigue and generalized weakness with any work and (41%) sometimes need to help from other to perform the sport activities.

Whereas a cross-sectional study done in Qatar by (Nashwan *et al*., 2018), which measure the QOL in 40 thalassemic adolescents and 40 healthy adolescents they found that the total average physical performance for all items such as walking, running, taking shower, lifting heavy things and doing sports & activities. This equated to (78.2) points, indicating that these teenagers were physically capable of functioning despite their sickness and health state.

The findings of physical health related to school in the present study as follows, two fifth of children always have school absenteeism because of tired, less than half of them sometimes Performing school home-work with difficulty due to fatigue, two fifth of them sometimes their attention drop during the lesson due to fatigue, and less than half of them always have exertion to go to the school-picnics. More supportive evidence is provided by (Kshaish & Aziz, 2020), which found the (51%) of thalassemic school age children sometimes have school absenteeism, (53%) sometimes effect school homework, (55%) sometimes, affect on their attention during lessons, (46%) sometime affect on their understanding to the lessons, and (56%) sometimes, has difficult going to school-picnic. The study done by (Nashwan *et al*., 2018), which find adolescents with thalassemia had also a significant decrease in school performance. Other study which directed by (Elzaree *et al*., 2018), they observed that thalassemia major has a bad and negative effect on the school commitment.

Regarding to physical health related to playing, the result of current study (table 4.4) shown more than two fifth never have efforts in playing with other children, more than half never find it hard to do their favorite hobbies and more than half always looking for activities that don't require a lot of energy. Researcher's point of view; the patients with hemoglobinopathies suffer from weakness and tiredness so the majority of these patients have difficulties and problems in the playing activities, but the present study finds the majority of patients always favorite or like to perform easy activities such as drawing or doing electronic games that don’t need a lot of energy so, the majority of the sample show never have effort in playing.

Kshaish & Aziz, (2020) noticed that the playing and hobbies in thalassemic school age children (52%) sometimes have difficulties in playing with other kids, (51%) sometimes have difficulties to play with their favorite hobbies and (47%) sometimes, like to find other hobbies that it does not require a lot of energy.

A cross sectional study design done by (Batool *et al*.,2017) conducted on 91 patients with thalassemia major ages 12 _ 18 years old, which revealed that Majority of the patients reported inability to play outdoor. (Akter *et al*.,2020), which they notice that majority of thalassemic children were unable to perform intensive physical activity They are unable to engage in normal activities such as homework, playing a variety of games, or engaging in other individual activities.

Concerning to physical health related to personal hygiene, the result of current study (table 4.4) shown more than three quarters of hemoglobinopathic adolescents never have difficulties in taking bath or shower alone, majority of them never find struggle in drying their bodies, more than half of them never need help from others to prepare hair style, most of them never require help by others in toilet training, majority of them never need help by others in fingernails care, and more than three quarters of them never have trouble in brushing teeth. Whereas, (Kshaish & Aziz, 2020) noticed that majority of thalassemic school age children have sometimes effect of personal hygiene activities.

This result could be rationalized to the majority of hemoglobinopathy adolescents in the present study had never effect on their body hygiene, this may be attributed to their ages as in majority aged 14-19; as this age is characterized by taking care of their appearance and cleanliness, especially since most of the sample is from the urban, not from the villages and the outskirts of Babylon, and it considered as consequences of their cultures, educational level, and awareness of their problems.

The findings of physical health related to sleep in the present study (table 4.4) shown more than one third of hemoglobinopathic adolescents always go to bed regularly and more than two fifth sometimes have distress in sleeping. Similar finding in study conducted by (Bagul *et al*.,2013) on (54) thalassemic children and controls above 9 years old, were studied (Psychosocial issues for adolescents and young adults with thalassemia) they found majority of patients had sleep problems. The results of study directed by (Kshaish & Aziz, 2020), they find (61%) of patients sometimes having regular sleep pattern and (51%) sometimes having sleep difficulty.

Many hemoglobinopathic patients, according to the result of the present study, may have sleep problems due to the nature of the disease such as extramedullary hematopoiesis in the upper airway area of the affected child, which could be a cause of obstructive sleep apnea. Furthermore, a lack of familial and environmental factors such as marital conflict, economic status, a suitable sleeping environment, reduced cognitive and educational performance, and rising psychological conditions such as behavioral issues such as symptoms of anxiety and depression may contribute to the sleep disturbance for the affected children's.

5.4: Discussion of the responses of psychological wellness for hemoglobinopathic adolescents

The present study reveals the overall findings of psychological wellness among hemoglobinopathy adolescents according to (table 4.7) which revealed that there are more than half of the sample have good psychological wellness, less than two-fifths have moderate Psychological Wellness, and a little bit of them has poor Psychological Wellness.

Similar results shown in the study conducted in Babil / Iraq, on (100) thalassemic and (100) healthy adolescents by (Saeed &Aldoori,2020), in order to identify the "Psycho-social Development of Thalassemic and Healthy Adolescents: Comparative Study" they found that psychological development for the half of adolescents with thalassemia have good response, while less than half of them is fair response, and little bit of them have poor response.

Other study which conducted in Al Najaf Al Ashraf / Iraq by (Al Ebrahimy *et al*.,2016), on (100) school age children with thalassemia in order to assess their psychosocial aspects, they found that the emotional domain of thalassemic children has the greatest impact on psychological features. This outcome is consistent with the findings of the present study in the psychological wellness for the hemoglobinopathic adolescents.

Further supported evidence is a cross-sectional study done by (Shafie *et al*.,2020), to assess HRQoL for (368) thalassemic children in Malaysia aged (3 _ to 18) years, which found that HRQoL has progressed in the last decade due to a better approach in treatment, whereas the enhancement of school functioning aspect is requiring additional efforts.

The researcher believes that adolescent response to psychological wellness within an acceptable limit may be due to they did not express to the reality of feelings because of their shyness or trying to escape from a weak sensation or coping with their illness status because of their cognitive awareness, a cultural issues, and religion, for example, one of the participants in the present study was a female aged 17 years said that she had suicidal thought and when she became to try doing of her idea she stopped because of her religion, and the small age patients will not be thinking about their emotions. Any way this refers to a defensive mechanism from adolescents to be better than others, but it is not a perfect response so should make all hemoglobinopathy children under monitoring for their psychological wellness to prevent relapse and controlling of the comorbid conditions such as anxiety and depression because of the affected children at high risk for psychological distress due to the nature of their chronic illness.

5.5. Discussion of the responses of social wellness for hemoglobinopathic adolescents

Analysis of such information in the present study (Table 4.9) shows overall social wellness for hemoglobinopathy adolescents that more than half of the sample has good social wellness, more than one-quarter of the Sample has moderate social wellness, and less than one-quarter of them have poor social wellness.

This result goes in line with the findings of study directed by (Saeed &Aldoori,2020), they found that more than half of both thalassemic and healthy adolescents highly response, also both of thalassemic and healthy adolescents showed less than half fair response, while no poor response to social developments in both them.

A cross-sectional study conducted on 40 thalassemic adolescents and 40 healthy adolescents, also agreed with the current study in social aspects, which aimed to measure the quality of life for adolescents aging (14_18 years) with major thalassemia in Qatar done by (Nashwan *et al*., 2018). Their results showed that patients with thalassemia had a lower quality of life score "(69.1±16.8)" than healthy matched children "(77±12.8), ($p < 0.001$)". In terms of social, physical, and emotional aspects, both of them were identical. The school's performance is also significantly poorer in thalassemic patients.

Another study conducted in Egypt, by (Hassan & Azzab, 2016), who found that the health instructions affect the quality of life and psychological problems among thalassemic children, and more than one-third of children with thalassemia reported having a good social relationship.

5.6: Discussion the correlation between physical, psychological, and social wellness

According to tables (4.10), (4.11), and (4.12) in the present study shows that a highly significant correlation between physical wellness, psychological wellness, and social wellness at P-value ≤ 0.05.

From the researcher's point of view, patients with hemoglobinopathies disorders had a highest risk of psychosocial distress such as stress, anxiety, depression, low self-esteem, lack of self-efficacy, and weakness of social relationships due to the nature of their illness, the frequency of their visits to the hospital, the large number of treatments used and the large number of blood transfusions, in addition to the negative impact of the disease on their physical forms and growth rate, and performance of their daily living activities. All of these causes which has negative effect on their wellness. This means that there is a close interrelationship between both physical and psychosocial wellness for hemoglobinopathic patients.

5.7: Discussion of the Association between physical, psychological, social wellness, and demographic data

The findings of current study table (4.13) indicated that there is a highly significant association between physical wellness and several demographical "data at $p \leq 0.05$", like (age, patient level of education, mother education, father education, parent consanguinity).

Similar result in descriptive cross-sectional study was conducted on 100 adolescents with hemolytic anemia by (Issa *et al*.,2018), in order to assess "Self-concept in Relation to Hemolytic Inheritance Blood Diseases among Adolescents in Babylon Governorate-Iraq" they found that adolescents age, mother education, and father education are highly significant correlation with low self-concept level among sample.

Researchers opinion that older children may have increased their abilities for doing their daily living activities independently, and the higher education level for both patients and their parents leads to an increase in their cognitive abilities and understanding of the disease and its treatment, which may lead to enhance their physical wellness.

Regarding to association between psychological wellness and demographic data, in the present study (table 4.14) demonstrates that there is a highly significant association between psychological wellness and some demographical data, such as (patient Level of education, Economic state of

Family). This result agree with the finding of study done by (Al Ebrahimy *et al*.,2016), they found that there is a strong significant link between psychosocial aspect for thalassemic school age children and their education level, socioeconomic state.

Concerning to association between social wellness and demographic data, current study at table (4.15) reveals that there is a highly significant associations between social Wellness and several demographical “data at $p \leq 0.05$”, like patient level of education, mothers’ occupation, economic state of family.

From the researcher's point of view, the decline of patients' level of education and their low socioeconomic status which had negatively effect on their psychosocial wellness. available supporting evidence in a case control study done by (Mettananda, 2020) on 288 transfusion thalassemic patients and similar controls were recruit for (Determination of psychological morbidity in pediatric with transfusion-dependent β-thalassemia and their parents in Srilanka) demonstrated that low family income had significant effect on their situation.

A cross-sectional analytical study directed by (Thiyagarajan *et al*.,2019), which conducted on 125 patients with thalassemia and 125 parents in order to (Assessing the role of family well-being on the QoL for thalassemic Indian children) they found that the three factors such as children education, family income, and parent education having significantly contributed to the children’s health-related quality of life among kids affected with disease.

Other cross-sectional study conducted by (Khamoushi *et al*.,2015), on 64 patients with major thalassemia for determine the (Stress, Anxiety, and Depression Prevalence and Socio-Demographic Characteristics among Patients with Thalassemia Major in Kermanshah County) which found that education level has significant relationship with depression, and patient with thalassemia major under high risk for developing of mental distress.

5.8: Discussion of the Association between physical, psychological, social wellness, and medical history

Table (4.16) in the current study show that there is a non-significant relationship between physical wellness and any of medical history at P-value ≤ 0.05. Whereas the association between psychosocial wellness and medical history according to tables (4.17) and (4.18) in the current study reveals that there is a highly significant relationship between psychosocial wellness and some medical history at $p \leq 0.05$, such as (Type of illness, Splenomegaly, Hepatomegaly).

The researcher believes that may be thalassemia major is more effective than other types of hemoglobinopathies disorders on psychosocial wellness for adolescents, because it is more widespread than other types in addition it more associated with the occurrence of the complications such as splenomegaly and hepatomegaly that increasing the suffering of patients.

Table (4.19) reveals that there is a significant correlation between hemoglobin and the three variables (physical, psychological and social) whereas a highly significance effected by hemoglobinopathies at P-value ≤ 0.05.

Mikael & Al-Allawi, (2018) they found in their study the hemoglobin level is one of the most important factors that effect on the quality of life for Iraqi Kurdistan adolescents and children.

The researcher's opinion is that the low hemoglobin level among the affected patients may be related to an increase in their fatigue and exhaustion, which lead to a negative impact on both physical and psychosocial wellness for hemoglobinopathy adolescents.

At the end of the study, the researcher found that the sample has many psychological and social health problems that can be due to the abnormal condition in the red blood cells, which leads to the shortage of hemoglobin, and in turn will effects on their quality of life and will be a burden on their peers and families.

CHAPTER SIX

CONCLUSIONS AND RECOMMENDATIONS

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CONCLUSIONS AND RECOMMENDATIOS

This chapter presents the conclusions which are derived from the interpretation and discussion of the study findings. The recommendations are based on the study outcomes.

6. 1. Conclusions:

The study reaches to the following conclusions based on the interpretation and discussion of the study findings.

1. Most of the hemoglobinopathic adolescents in the study sample are mean ages of 14.7 years, and are evenly distributed by gender, with a majority of participants from rural areas.

2. Majority of study sample's and their families had a primary level of education, who feel sufficient to some extent in their socio-economic status in the majority of hemoglobinopathic patients' families, which leads them to have more than one child effected with this genetic disease.

3. Hemoglobinopathic disorders had an impact on the physical, psychological and social wellness of the patients.

4. The majority of affected adolescents experience complication and most of them had splenomegaly, and the BMI percentile for less than three quarters of them between 5th to less than 85th percentile.

5. Age of children and their educational status as well as parent educational level, parent consanguinity, and hemoglobin level had significant impact on the physical wellness of the study sample.

6. The patients and their parent’s educational level, socio-economic status, hemoglobin level and type of illness, has a great effect on the overall psychosocial wellness of the study sample.

6.2. Recommendations

Based on the early stated conclusion, the present study can recommends that:

1. Pediatric nurses and other health care workers must be attentive about the need of determining physical and psychosocial wellness in children with hemoglobinopathic disorders in order to modify effective management as early detection, complication prevention.

2. Activate the roles related of pediatric nurses and other health workers in promoting and encouraging children with hemoglobinopathic disorders to achieve improved performance in their daily living activities, psychological wellness and social relationship, Particularly for children with little social support and a high severity of sickness. The most important thing is to include their families in caring for them to increase their awareness of taking care of them in all aspects that affect and weaken their personalities.

3. Encourage health promoting programs to enhance wellness and prevent comorbid illness that occur with these disorders such as depression and anxiety. As well as encourage the limitation of the consanguinity marriage which lead to control of the prevalence of the genetic disease.

4. Encourage nurses to make good documentation program for register patients in the center and inform ministry of health for necessary increasing the number of centers in the province to decrease load, promote management and encourage patients from far resident to follow up easily, by setting incentives to adhere to the appointments, and most likely to help them reduce or raise the cost of treatment and the device needed to take treatment.

5. In the hereditary blood disease center , psychologists and sociologists are needed to offer a link between patients, schools, families, and health care provider.

6. Teachers in primary, intermediate, and secondary schools who are responsible for educating pupils and supporting them psychosocially should attend training classes. These courses may be from responsibility of the primary health care providers activities.

7. To improve care and alleviate the suffering of hemoglobinopathies adolescents, thorough continuous comprehensive support is required, as well as surveying their health and medical state.

8. It is recommended that further studies be conducted to assess the physical and psychosocial wellness for different age groups and a larger sample that could include all centers in the country.

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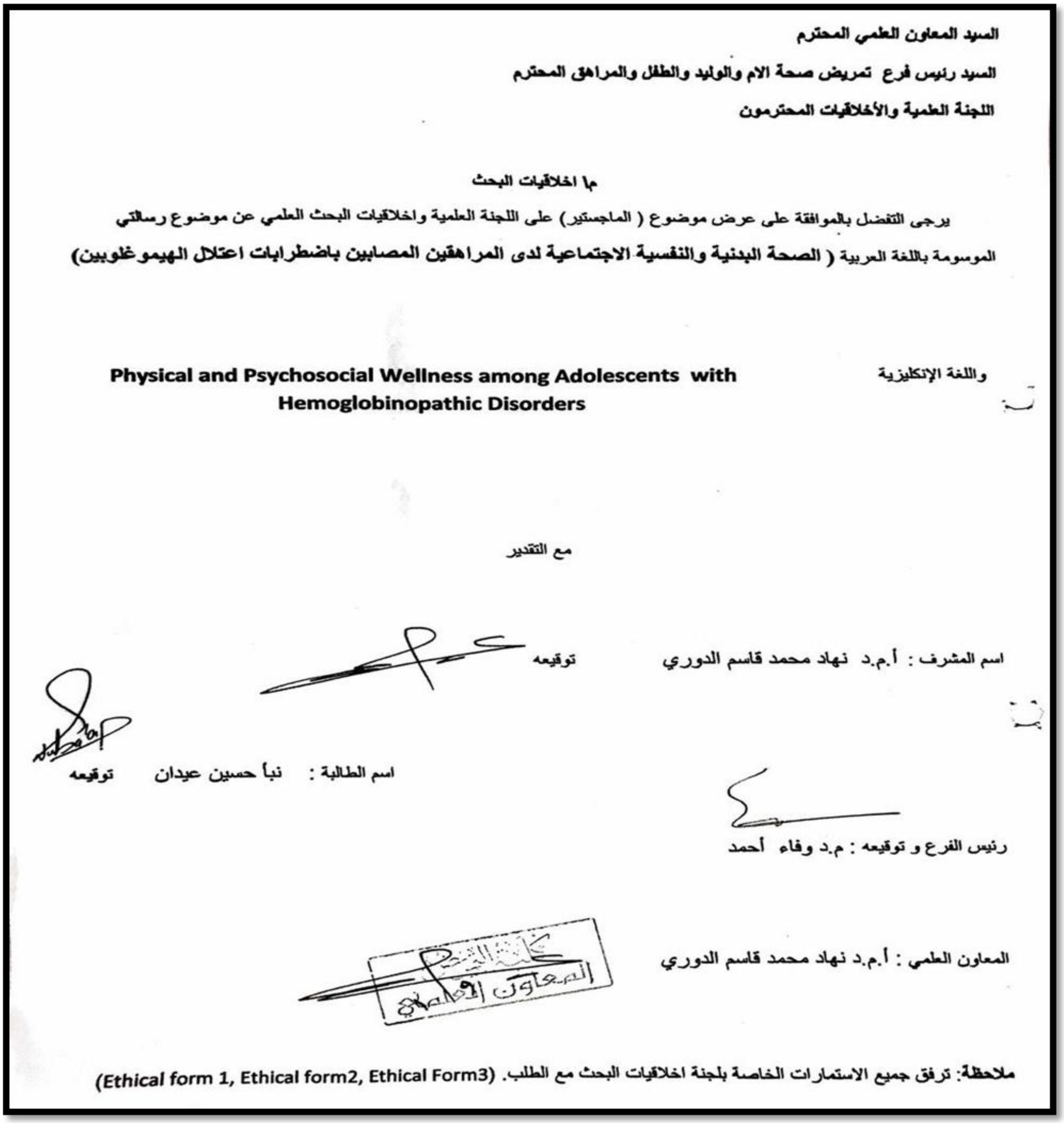
APPENDICES

APPENDIX (A)

ADDEMINSTRIVE ARRANGEMENTS



السيد المعاون العلمي المحترم

السيد رئيس فرع تمريض صحة الام والوليد والطفل والمراهق المحترم

اللجنة العلمية والأخلاقيات المحترمون

م/ اخلاقيات البحث

يرجى التفضل بالموافقة على عرض موضوع (الماجستير) على اللجنة العلمية واخلاقيات البحث العلمي عن موضوع رسالتي الموسومة باللغة العربية (الصحة البدنية والنفسية الاجتماعية لدى المراهقين المصابين باضطرابات اعتلال الهيموغلوبين)

واللغة الإنكليزية **Physical and Psychosocial Wellness among Adolescents with Hemoglobinopathic Disorders**

مع التقدير

اسم المشرف : أ.م.د نهاد محمد قاسم الدوري توقيعه

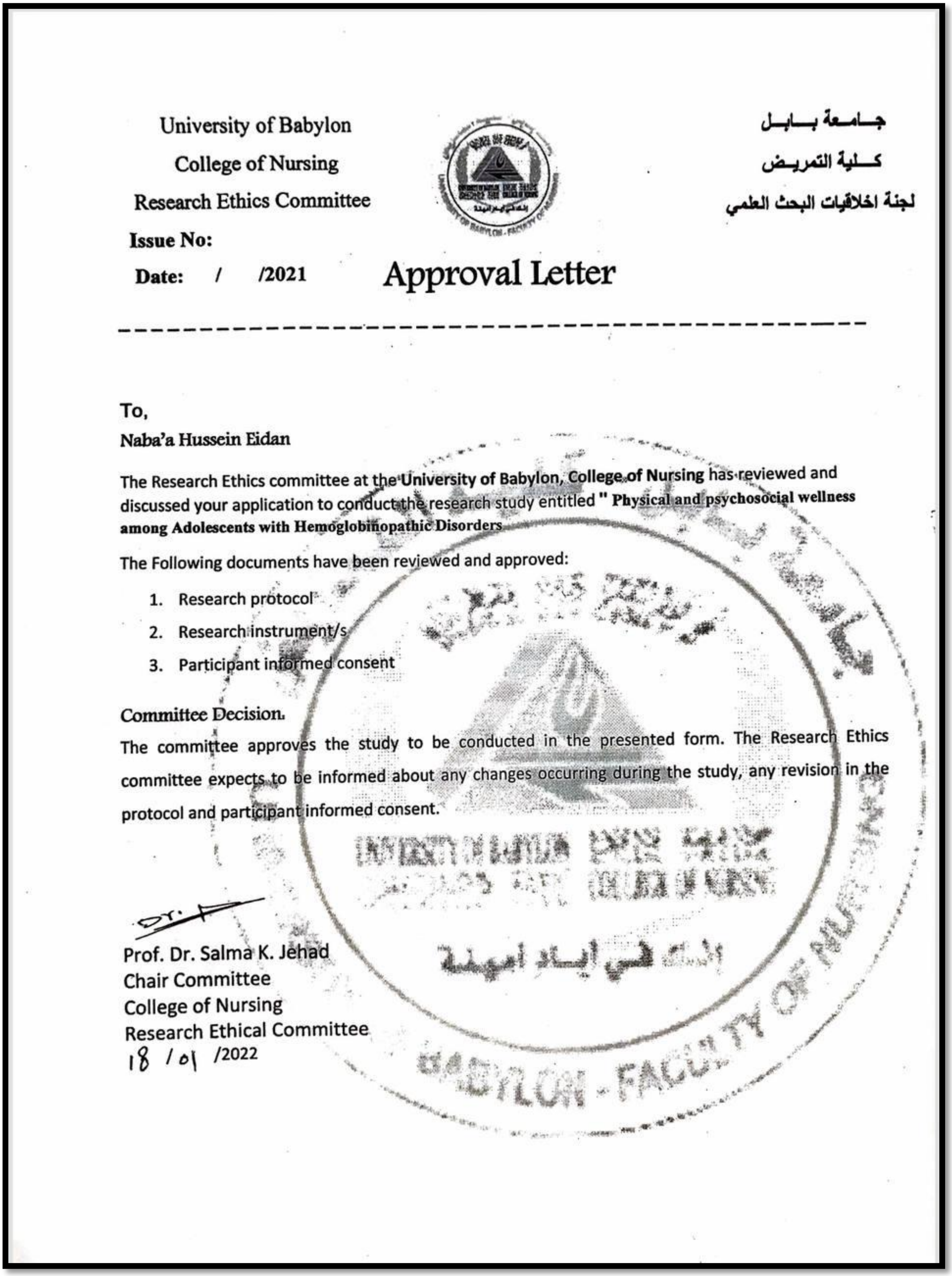
اسم الطالبة : نبأ حسين عيدان توقيعه

رئيس الفرع و توقيعه : م.د وفاء أحمد

المعاون العلمي : أ.م.د نهاد محمد قاسم الدوري

كلية التمريض
المعاون العلمي

ملاحظة: ترفق جميع الاستمارات الخاصة بلجنة اخلاقيات البحث مع الطلب. (*Ethical* form 1, Ethical form2, Ethical Form3)



University of Babylon
College of Nursing
Research Ethics Committee

جـامـعة بـابـل
كـلية التمريـض
لجنة اخلاقيات البحث العلمي

Issue No:

Date: / /2021

Approval Letter

To,

Naba'a Hussein Eidan

The Research Ethics committee at the **University of Babylon, College of Nursing** has reviewed and discussed your application to conduct the research study entitled **" Physical and psychosocial wellness among Adolescents with Hemoglobinopathic Disorders**

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
18 /01 /2022

APPENDICIES

Ministry of Higher Education and Scientific Research

جمهورية العراق

وزارة التعليم العالي والبحث العلمي

University of Babylon
College of Nursing

جامعة بابل
كلية التمريض
لجنة الدراسات العليا

العدد : ٤٩٧
التاريخ : ٢٧/ ١ /٢٠٢٢

Ref. No. :
Date: / /

الى / دائرة صحة بابل / مركز التدريب والتطوير
م/ تسهيل مهمة

تحية طيبة :

يطيب لنا حسن التواصل معكم ويرجى تفضلكم بتسهيل مهمة طالبة الماجستير (نبأ حسين عيدان) لغرض جمع عينة دراسة الماجستير والخاصة بالبحث الموسوم :

الصحة البدنية والنفسية الاجتماعية لدى المراهقين المصابين باضطرابات اعتلال الهيموغلوبين.

Physical and Psychosocial wellness among Adolescents with Hemoglobinopathic Disorders.

مع الاحترام ...

المرافقات //
- بروتوكول.
- استبانة.

ا.م . د. نهاد محمد قاسم الدوري
معاون العميد للشؤون العلمية والدراسات العليا
٢٠٢٢/١/٢٧

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

E-mail:nursing@uobabylon.edu.iq

STARS

وطني 07711632208
المكتب 009647711632208

www.uobabylon.edu.iq

جمهورية العراق

وزارة الصحة والبيئة
دائرة صحة محافظة بابـــــل
المديـــر العـــــام
مركـــز التدريب والتنمية البشرية
وحدة أدارة البحوث

Ministry Of Health
Babylon Health Directorate
Email:-
Babel_Healthmoh@yahoo.com
Tel:282628 or 282621

العدد : ١٢١
التاريخ : ١ / ٢ / ٢٠٢٢

وزارة الصحة
دائرة صحة بابل
مركز التدريب والتنمية البشرية

إلى / مستشفى الامام الصادق (ع)
مستشفى بابل للنسائية والأطفال
م/ تسهيل مهمة

السلام عليكم ...

أشارة إلى كتاب جامعة بابل /كلية التمريض / لجنة الدراسات العليا ذي العدد ٤٩٧ في ٢٠٢٢/١/٢٧

نرفق لكم ربطا استمارات الموافقة المبدئية لمشروع البحث العائد للباحثة طالبة الماجستير (نبأ حسين عيدان)

للتفضل بالاطلاع وتسهيل مهمة الموما أليه من خلال توقيع وختم استمارات أجراء البحث المرفقة في مؤسساتكم وحسب الضوابط والإمكانات لاستحصال الموافقة المبدئية ليتسنى لنا أجراء اللازم على أن لا تتحمل مؤسساتكم أية تبعات مادية وقانونية .... مع الاحترام

السيد المدير المحترم
لا مانع لدينا [illegible] مع الاحترام

د. ثبات ريس الفيحان
وحدة التعليم [illegible]
مستشفى بابل للنسائية والأطفال

المرفقات :
استمارة عدد /٢

[illegible]

الدكتور
محمد عبد الله عجرش
م/مدير مركز التدريب والتنمية البشرية
٢٠٢٢/ /

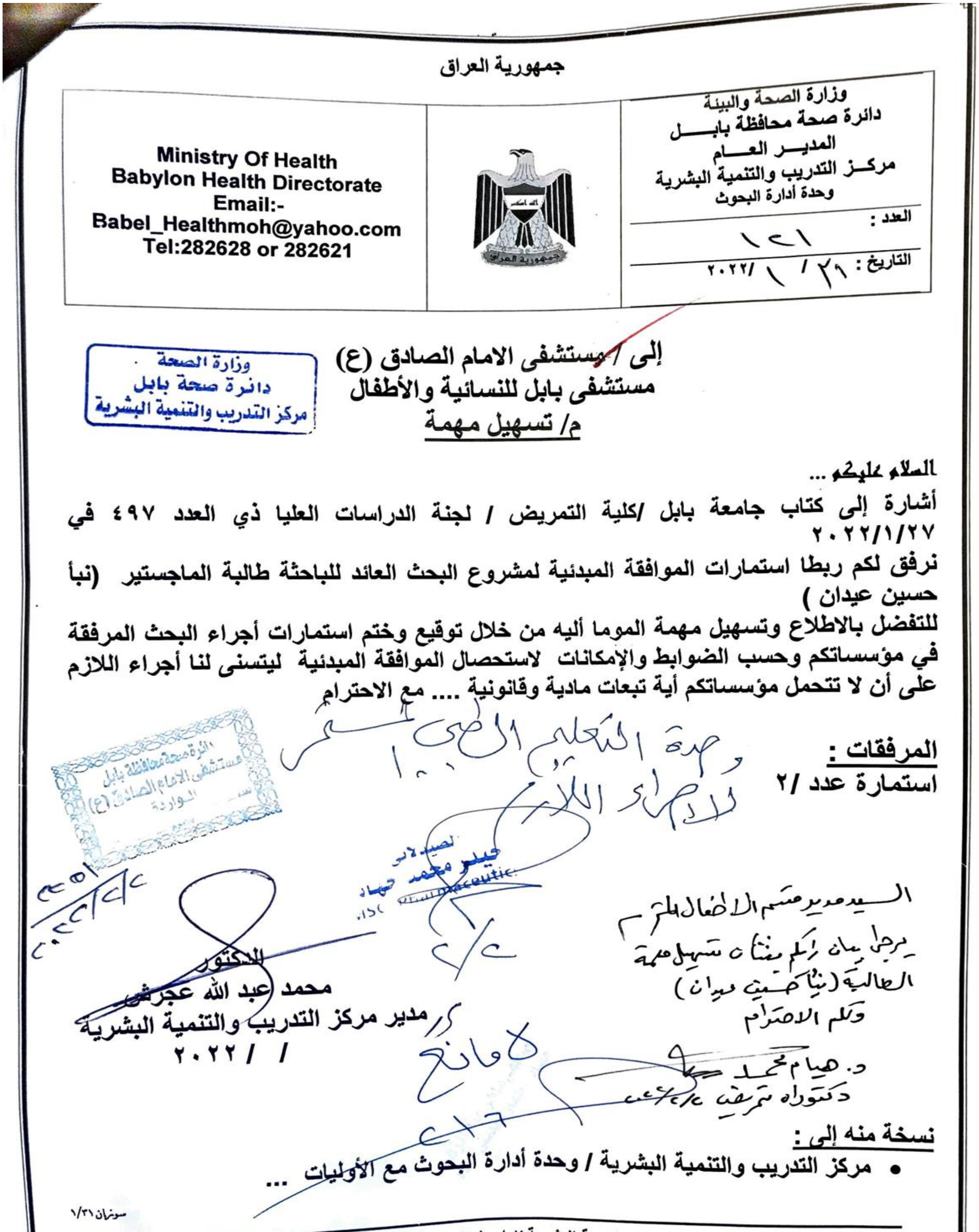
الدكتورة
زهراء جاسم السلطاني
اختصاص طب طوارئ
٢/ ٢/ ٢٠٢٢

نسخة منه إلى :

- مركز التدريب والتنمية البشرية / وحدة أدارة البحوث مع الأوليات ...

سوزان ١/٣١

دائرة صحة محافظة بابل / مركز التدريب والتنمية البشرية // ايميل المركز babiltrainning@gmail.com



جمهورية العراق

وزارة الصحة والبيئة
دائرة صحة محافظة بابل
المدير العام
مركز التدريب والتنمية البشرية
وحدة أدارة البحوث

العدد : ١٢١

التاريخ : ٢٩ / ١ / ٢٠٢٢

Ministry Of Health
Babylon Health Directorate
Email:-
Babel_Healthmoh@yahoo.com
Tel:282628 or 282621

وزارة الصحة
دائرة صحة بابل
مركز التدريب والتنمية البشرية

إلى / مستشفى الامام الصادق (ع)
مستشفى بابل للنسائية والأطفال
م/ تسهيل مهمة

السلام عليكم ...

أشارة إلى كتاب جامعة بابل /كلية التمريض / لجنة الدراسات العليا ذي العدد ٤٩٧ في ٢٠٢٢/١/٢٧

نرفق لكم ربطا استمارات الموافقة المبدئية لمشروع البحث العائد للباحثة طالبة الماجستير (نبأ حسين عيدان)

للتفضل بالاطلاع وتسهيل مهمة الموما أليه من خلال توقيع وختم استمارات أجراء البحث المرفقة في مؤسساتكم وحسب الضوابط والإمكانات لاستحصال الموافقة المبدئية ليتسنى لنا أجراء اللازم على أن لا تتحمل مؤسساتكم أية تبعات مادية وقانونية .... مع الاحترام

وحدة التعليم الطبي المستمر
للإجراء اللازم
١/٣١

دائرة صحة محافظة بابل
مستشفى الامام الصادق (ع)
الواردة

المرفقات :
استمارة عدد /٢

الصيدلاني
حيدر محمد جهاد

٢٠٢٢/٢/٢

٢/٢

الدكتور
محمد عبد الله عجرش
مدير مركز التدريب والتنمية البشرية
/ / ٢٠٢٢

السيد مدير قسم الأطفال المحترم
يرجى بيان رأيكم في تسهيل مهمة
الطالبة (نبأ حسين عيدان)
ولكم الاحترام
د. هيام محمد
دكتوراه تمريض ٢٠٢٢/٢/٢

لا مانع
٢/٦

نسخة منه إلى :

- **مركز التدريب والتنمية البشرية / وحدة أدارة البحوث مع الأوليات ...**

سوزان ١/٣١

دائرة صحة محافظة بابل / مركز التدريب والتنمية البشرية // ايميل المركز babiltrainning@gmail.com

جمهورية العــــراق

Ministry Of Health
Babylon Health Directorate
Email:-
Babel_Healthmoh@yahoo.com
Tel:282628 or 282621

وزارة الصحة والبيئة
دائرة صحة محافظة بابـــــل
المديـــر العــــام
مركـــز التدريب والتنمية البشرية
لجنة البحوث

استمارة رقم :- ٢٠٢١/٠٣

وزارة الصحة
دائرة صحة بابل
مركز التدريب والتنمية البشرية
لجنة البحوث

رقم القرار :- ٢٧
تاريخ القرار :-٤/ ٢ /٢٠٢٢

قرار لجنة البحوث

تحية طيبة ...

درست لجنة البحوث في دائرة صحة بابل مشروع البحث ذي الــرقم (٢٠٢١/٠١٨/ بابل) المعنون (الصحة البدنية والنفسية الاجتماعية لدى المراهقين المصابين باضطرابات اعتلال الهيموغلوبين) والمقدم مــن الباحثة (نبأ حسين عيدان) إلى وحدة أدارة البحوث والمعرفي مركز التدريب والتنمية البشرية في دائرة صحة بابل بتاريخ ٢٠٢٢/٢/٧ وقررت :

قبول مشروع البحث أعلاه كونه مستوفيا للمعايير المعتمدة في وزارة الصحة والخاصة بتنفيذ البحوث ولا مانع من تنفيذه في مؤسسات الدائرة .

مـــــــع الاحتـــــرام

الدكتور / محمد عبد الله عجرش
رئيس لجنة البحوث
٢٠٢٢/ /

نسخة منه إلى :

- مكتب المدير العام / مركز التدريب والتنمية البشرية / وحدة إدارة البحوث ... مع الأوليات.

سوزان

دائرة صحة محافظة بابل / مركز التدريب والتنمية البشرية // ايميل المركز babiltrainning@gmail.com

Appendix (B)

Questionnaire

Part I: Socio-demographic Information

1. **Age** ☐
2. **Gender**

   Male ☐ **Female** ☐

3. **patient Order among Siblings** ☐

4. **patient Level of Education**

   Unable to read and write ☐ Primary level ☐

   Intermediate level ☐ Secondary level ☐

5. **Mother Education**

   Unable to read and write ☐ Primary level ☐

   Secondary level ☐ Diploma or above ☐

6. **Father Education**

   Unable to read and write ☐ Primary level ☐

   Secondary level ☐ Diploma and above ☐

7. **Mothers' Occupation**

   Employed ☐ Unemployed ☐

8. Fathers' Occupation

Employed ☐ Unemployed ☐

9. Parent Consanguinity:

Yes ☐ No ☐

10. Economic State of Family:

Enough ☐ Enough to some extent ☐ Not enough ☐

11. Residency

Urban ☐ Rural ☐

Part II: Medical History

1. Type of illness

Major Thalassemia ☐ Intermediate thalassemia ☐

Sickle Beta Thalassemia ☐ Sickle cell anemia ☐

2. Frequency of blood transfusion/ month

One time ☐ Twice ☐ Three times & over ☐

3. Hemoglobin level ☐

4. Age at diagnosis: ☐

5. Disease complication

Osteoporosis ☐ yes ☐ no
Splenomegaly ☐ yes ☐ no
Hepatomegaly ☐ yes ☐ no
Hepatitis ☐ yes ☐ no
Diabetes ☐ yes ☐ no
Cardiomegaly ☐ yes ☐ no

6. Patient Weight ☐ Height ☐

7. Is there more than one affected child in the family

Yes ☐ No ☐

Part III: Physical Wellness for Hemoglobinopathy Adolescents

list	Item	Always	Sometime	Never
1	Have difficulties in wearing my clothes			
2	Need help from other to keep and arrange my clothes			
3	Have difficulties to care of my general appearance			
4	Have difficulty in walking alone			
5	Lift the heavy things with difficulty			
6	Have efforts in going to school			
7	Have fatigue and generalized weakness			
8	Need help from other to perform the sport activities			

9	Have school absenteeism because of tired			
10	Perform my school home-work with difficulty due to fatigue			
11	My attention drops during the lesson due to fatigue			
12	Have exertion to go to the school-picnics			
13	Have e efforts in playing with other children			
14	Find it hard to do my favorite hobbies			
15	I'm looking for other activities that don't require a lot of energy			
16	Have difficulties in taking bath or shower by my self			
17	Find struggle in drying my body			
18	Need help from others to prepare my hair style			
19	Require help by others in toilet training			
20	Need help by others in fingernails care			
21	I have trouble in brushing teeth			
22	Go to bed regularly			
23	Have distress in sleeping			

Part IV: Psychological Wellness for Hemoglobinopathy Adolescents

List	Item	Always	Sometime	Never
1	Suppose perfect well even in good health			
2	Think in need for good tonic			
3	Feel tired and exhausted			
4	Feel sick			
5	Suffer from a headache			
6	Feel tightness and pressure in my head			
7	Feel sudden states of cold or heat			
8	I have decrease in my sleeping hours due to anxiety			
9	If I fall asleep, I find it hard to stay asleep			
10	Feel tired all the time			
11	Get nervous quickly and in a bad mood			
12	Feel scared or panicky for no good reason			
13	Feel like I carry the worries of the world on My head			
14	Feel nervous and strung-up all the time			
15	Feel like I can finish myself with certain things			
16	Taking longer over the things I do			
17	Feel on the whole I'm doing things well			
18	I am satisfied with the way I’ve carried out my task			
19	Feel playing a useful part in things			
20	Can make decisions			
21	I am able to enjoy my normal day-to-day activities			
22	Feel like a worthless person			
23	Felt that life is entirely hopeless			
24	Feel that life is not worth living			

25	Have the idea of excluding my life			
26	Found at times I couldn't do anything because my nerves were too bad			
27	Wish I was dead and away from everything Around me			
28	Find the idea of getting rid of my life on my mind a lot			
29	Have trouble in understanding my lessons			
30	I am distressed in coping in school and other Pupils			
31	Have difficulties in understanding my administration about my health status			
32	Achieve low grades in school-examinations			

Part V: Social Wellness for Hemoglobinopathy Adolescents

List	**Item**	**Always**	**Sometime**	**Never**
1	Get emotional help and support I need from my family			
2	My family is willing to help me make decisions			
3	Can talk about my problem with my family			
4	My family support me to participate in social activities			
5	Get the same attention from my father and mother			
6	There is special person in my life who care about my feelings			
7	Find it difficult to make friends			
8	Find myself isolated among people			
9	My illness affects my relationship with my family and friends			
10	Suffer from a lack of adaptation to disease and society			

11	Find it difficult to speak in front of others			
12	My parents don’t listen to me when I talk			

Appendix (B)

استمارة استبيان

عزيزي /عزيزتي بين يديك استمارة استبيان بحث بعنوان **(الصحة البدنية والنفسية الاجتماعية لدى المراهقين المصابين باضطرابات اعتلال الهيموغلوبين)** لذا نلتمس عنايتكم في ملئ الاستمارة، كما نود اعلامكم انه لا توجد اجابة صائبة او خاطئة بل المقصود فيها التعبير الصادق عن نفسك. ارجو تفضلكم بالإجابة عنها جميعا ومن غير ترك اي فقرة منها ولا داعي لذكر الاسم، واحيطكم علما ان المعلومات التي ستذكر فيها، ستعامل بسرية تامة وتستخدم لأغراض البحث العلمي فقط.

اقرأ جيدا كل جملة ثم اجب عنها بوضع اشارة (√) تحت كلمة (دائما) اذا كانت العبارة تصف ما تشعر به عادة.

او تضع اشارة (√) تحت كلمة (احيانا) اذا كانت العبارة تصف ما تشعر به في بعض الاحيان.

او تضع اشارة (√) تحت كلمة (ابدا) اذا كانت العبارة لا تصف ما تشعر به عادة.

مثال

العبارة	دائما	احيانا	ابدا
اشعر باني تعبان ومرهق	√		

فإنني اضع اشارة (√) تحت دائما لان الجملة عبرت عما اشعر عادة

العبارة	دائما	احيانا	ابدا
اشعر باني تعبان ومرهق		√	

فإنني اضع اشارة (√) تحت احيانا لان الجملة عبرت عما اشعر به في بعض الاحيان

العبارة	دائما	احيانا	ابدا
اشعر باني تعبان ومرهق			√

اما اذا لم تعبر الجملة عما اشعر به عادة فإنني اضع اشارة(√) تحت ابدا

شاكرين تعاونكم معنا
الباحثة/ نبأ حسين عيدان
جامعة بابل/ كلية التمريض

الجزء الاول: المعلومات الديموغرافية للمريض والعائلة

1. **عمر المريض:** ☐

2. **الجنس :** ذكر ☐
انثى ☐

3. **تسلسل المريض بين اخوته :** ☐

4. **المستوى التعليمي للمريض:**
لا يقرأ ولا يكتب ☐ ابتدائية ☐
متوسطة ☐ اعدادية ☐

5. **المستوى التعليمي للام :**
لا تقرا ولا تكتب ☐ ابتدائية ☐
متوسطة او اعلى ☐ معهد او اعلى ☐

6. **المستوى التعليمي للاب:**
لا يقرأ ولا يكتب ☐ ابتدائية ☐
متوسطة او اعلى ☐ معهد او اعلى ☐

7. **عمل الام :** تعمل ☐ لا تعمل ☐

8. **عمل الاب :** يعمل ☐ لا يعمل ☐

9. **هل توجد صلة قرابة بين الوالدين:** نعم ☐ **لا** ☐

10. **الدخل المعيشي للأسرة:**
كافي ☐ يكفي الى حد ما ☐ غير كافي ☐

11. **محل السكن :** المناطق الريفية ☐ المنا طق الحضرية ☐

الجزء الثانى : التاريخ الطبى للمريض

1. **نوع التشخيص الطبي للمريض :**

ثلاسيميا كبرى ☐ ثلاسيميا متوسطة ☐

الثلاسيميا المنجلية ☐ فقر الدم المنجلي ☐

2. **عدد مرات نقل الدم شهريا:**

مره واحدة ☐ مرتان ☐ ثلاث مرات او اكثر ☐

3. **مستوى الهيموغلوبين** : ☐

4. **عمر الطفل عند التشخيص** : ☐

5. **مضاعفات المرض:**

هشاشة العظام نعم ☐ لا ☐

تضخم الطحال نعم ☐ لا ☐

تضخم الكبد نعم ☐ لا ☐

التهاب الكبد الفيروسي نعم ☐ لا ☐

السكري نعم ☐ لا ☐

امراض القلب نعم ☐ لا ☐

6. **وزن المريض** ☐ **طوله** ☐

7. **هل هناك اكثر من طفل مصاب بالمرض في العائلة** : نعم ☐ لا ☐

الجزء الثالث : الصحة البدنية للمراهقين المصابين باعتلال الهيموغلوبين

ت	الفقرة	دائما	احيانا	ابدا
1	ارتدي ملابسي بصعوبة			
2	أحتاج إلى مساعدة الآخرين للحفاظ على ملابسي وترتيبها			
3	أجد صعوبة في الاهتمام بمظهري العام			
4	اعاني من صعوبة المشي لوحدي			
5	أحمل الأشياء الثقيلة بصعوبة			
6	لدي صعوبات في الذهاب إلى المدرسة			
7	أعاني من إرهاق وضعف عام			
8	أحتاج إلى مساعدة الآخرين لأداء الأنشطة الرياضية			
9	اتغيب عن المدرسة بسبب التعب			
10	أؤدي واجباتي المدرسية بصعوبة بسبب الارهاق			
11	انتباهي ينخفض خلال الدرس بسبب التعب			
12	لدي مشكلة في المشاركة بالفعاليات المدرسية			
13	اجد صعوبة في اللعب مع الأطفال الآخرين			
14	أجد صعوبة في ممارسة هواياتي المفضلة			
15	أحب أن أجد هوايات أخرى لا تحتاج إلى الكثير من الطاقة			
16	أجد صعوبة في الاستحمام بمفردي			
17	أجد صعوبة في تجفيف جسدي			
18	أحتاج إلى مساعدة الآخرين في إعداد تسريحة وتصفيف شعري			
19	أحتاج إلى مساعدة الآخرين في استخدام المرحاض			
20	أحتاج إلى مساعدة الآخرين في العناية بأظافري			
21	لدي صعوبات في تنظيف الأسنان بالفرشاة			
22	أذهب إلى الفراش بانتظام			
23	لدي صعوبات في النوم			

الجزء الرابع: الصحة النفسية للمراهقين المصابين باعتلال الهيموغلوبين

ت	الفقرة	دائما	احيانا	ابدا
1	اشعر اني بخير وان صحتي جيدة			
2	اعتقد اني بحاجة الى ادوية مقوية			
3	اشعر باني تعبان ومرهق			
4	اشعر بالمرض			
5	اعاني من الام بالراس			
6	اشعر بشد وضغط في راسي			
7	اشعر بحالات مفاجئة من البرودة او السخونة			
8	لقد نقصت ساعات نومي بسبب القلق			
9	اذا غلبني النوم اجد صعوبة في ان اظل نائما			
10	اشعر باني مجهد بشكل دائم			
11	اتوتر بسرعة ومزاجي متعكر			
12	اشعر بالخوف او الرعب بدون سبب وجيه			
13	اشعر باني احمل هموم الدنيا على اكتافي			
14	اشعر بالغضب والتوتر طوال الوقت			
15	اشعر باني قادر على اشغال نفسي بأشياء معينة			
16	استغرق وقت طويل لغرض انجاز اعمالي			
17	اشعر بشكل عام باني اجيد ما أفعله			
18	اشعر بالرضا عن المستوى الذي اؤدي فيه اعمالي			
19	اشعر باني اقوم بدور مفيد في انجاز اعمالي			
20	استطيع اتخاذ القرارات			
21	استطيع الاستمتاع بنشاطاتي اليومية الاعتيادية			
22	اظن باني شخص بدون قيمة			
23	اعتقد بانه لا امل في الحياة على الاطلاق			
24	اشعر بان الحياة لا تستحق ان نعيشها			
25	تراودني فكرة التخلص من حياتي			
26	اشعر في بعض الاوقات بعدم استطاعتي على فعل اي شيء لان اعصابي متوترة			
27	اتمنى ان اكون ميتا وبعيد عن كل ما هو حولي			
28	اجد فكرة التخلص من حياتي تراود ذهني كثيرا			
29	اجد صعوبة في فهم دروسي			
30	اجد صعوبة في التأقلم مع المدرسة والتلاميذ الاخرين			
31	ادارة المدرسة لا تتفهم حالتي الصحية			
32	درجاتي منخفضة في الامتحانات المدرسية			

الجزء الخامس : الصحة الاجتماعية للمراهقين المصابين باعتلال الهيموغلوبين

ت	الصحة الاجتماعية	دائما	احيانا	ابدا
1	احصل على المساعدة والدعم العاطفي من عائلتي			
2	عائلتي مستعدة لمساعدتي في اتخاذ القرارات			
3	يمكنني التحدث عن مشاكلي مع عائلتي و اصدقائي			
4	عائلتي تدعمني للمشاركة في الانشطة الاجتماعية			
5	احصل على نفس الاهتمام من والدي ووالدتي			
6	هناك شخص مميز في حياتي يهتم بمشاعري			
7	اجد صعوبة في تكوين الاصدقاء			
8	اجد نفسي منعزلا بين الناس			
9	مرضي يؤثر على علاقتي بأفراد عائلتي والاصدقاء			
10	اعاني من عدم التكيف مع المجتمع			
11	اجد صعوبة في التحدث امام الاخرين			
12	والداي لا يستمعان الي عندما اتحدث			

APPENDIX (C)

PANEL OF EXPERTS

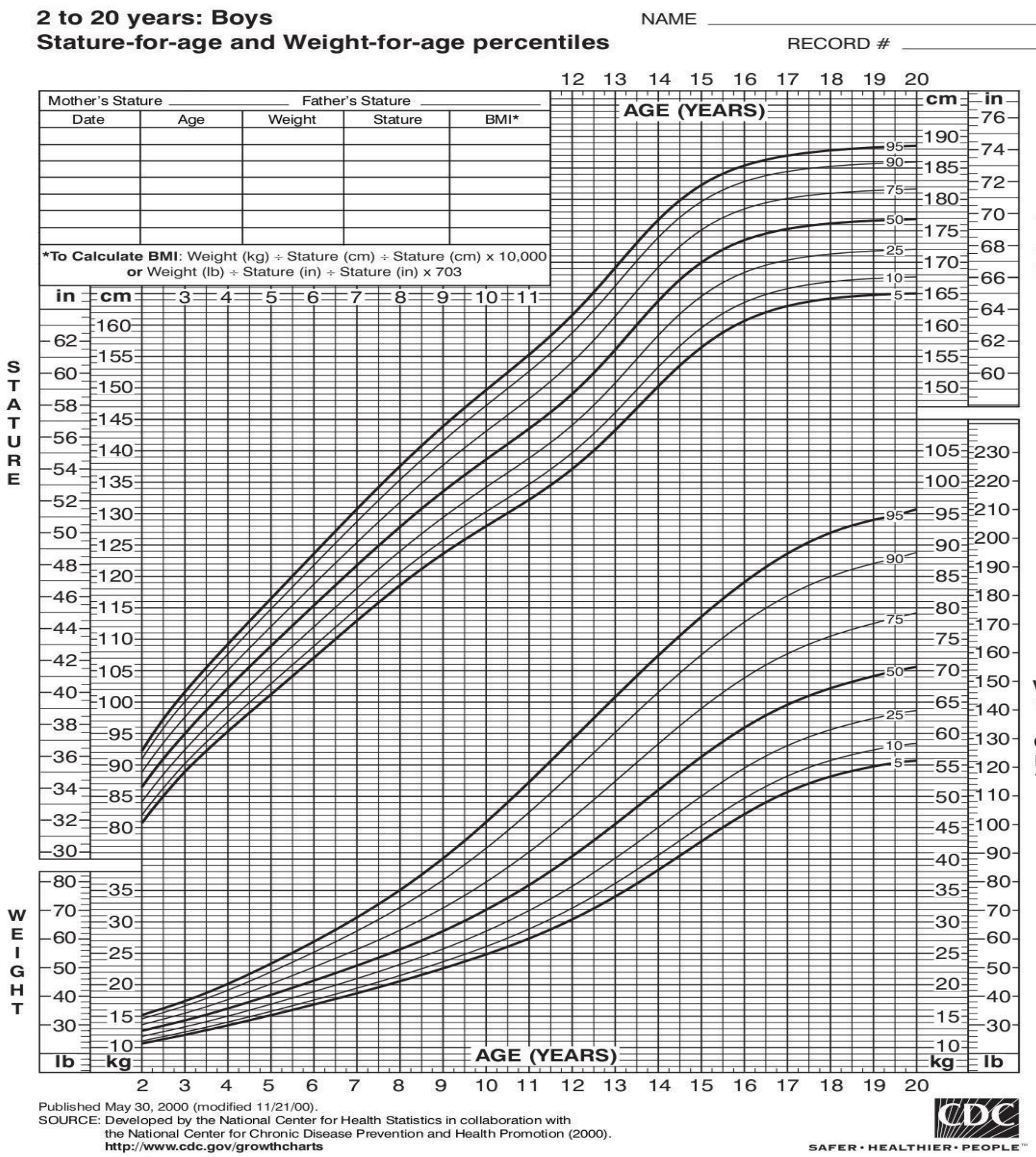
قائمة بأسماء الخبراء:

ت	اسم الخبير	الاختصاص	اللقب العلمي	مكان العمل	سنوات الخبرة
1	د. أمين عجيل الياسري	تمريض صحة الاسرة والمجتمع	استاذ	جامعة بابل/ كلية التمريض	37
2	د. سجاء هاشم محمد	تمريض الصحة النفسية والعقلية	استاذ	استاذ متمرس/ جامعة بابل/ كلية التمريض	40
3	د. عفيفة رضا عزيز	تمريض صحة الطفل والمراهق	استاذ	جامعة بغداد/ كلية التمريض	40
4	د. سلمى كاظم جهاد	تمريض صحة المجتمع	استاذ	جامعة بابل/ كلية التمريض	37
5	د. يحيى عبد الشهيد	طب اطفال	استاذ	جامعة بابل/ كلية الطب	36
6	د .عبد المهدي عبد الرضا	تمريض الصحة النفسية والعقلية	استاذ	جامعة بابل/ كلية التمريض	43
7	د. خميس بندر عبيد	تمريض الاطفال	استاذ	جامعة كربلاء/ كلية التمريض	21
8	د. محمد باقر حسن آل دخيل	تمريض الاطفال	استاذ مساعد	جامعة الكوفة/ كلية التمريض	18
9	د. حيدر حمزه علي	تمريض الصحة النفسية والعقلية	استاذ مساعد	جامعة الكوفة/ كلية التمريض	12
10	د. صافي داخل نوام	تمريض الصحة النفسية والعقلية	استاذ مساعد	جامعة كربلاء/ كلية التمريض	15
11	د. عذراء حسين شوق	تمريض الاطفال	استاذ مساعد	جامعة بغداد/ كلية التمريض	17
12	د. أحمد عبدالله عبد الحسيناوي	تمريض الاطفال	استاذ مساعد	جامعة ذي قار/ كلية التمريض	13
13	د. ناجي ياسر سعدون	تمريض صحة مجتمع	استاذ مساعد	جامعة بابل / كلية التمريض	33
14	د. ماهر خضير	اللغة العربية	استاذ مساعد	جامعة بابل / كلية التمريض	15
15	د. حسن علي حسين	تمريض الصحة النفسية والعقلية	استاذ مساعد	جامعة بغداد/ كلية التمريض	20

16	د. وفاء أحمد أمين	دكتوراه تمريض صحة الام والوليد	استاذ مساعد	جامعة بابل/ كلية التمريض	15
17	د. قحطان قاسم محمد	تمريض الصحة النفسية والعقلية	استاذ مساعد	جامعة بغداد / كلية التمريض	14
18	د. علي احمد كاظم الحطاب	تمريض الصحة النفسية والعقلية	مدرس	جامعة بابل / كلية التمريض	11
19	د. أحمد عبد المحسن فخري	طب الاطفال	مدرس	جامعة بابل /كلية الطب	10

APPENDIX (D)

GROWTH CHART

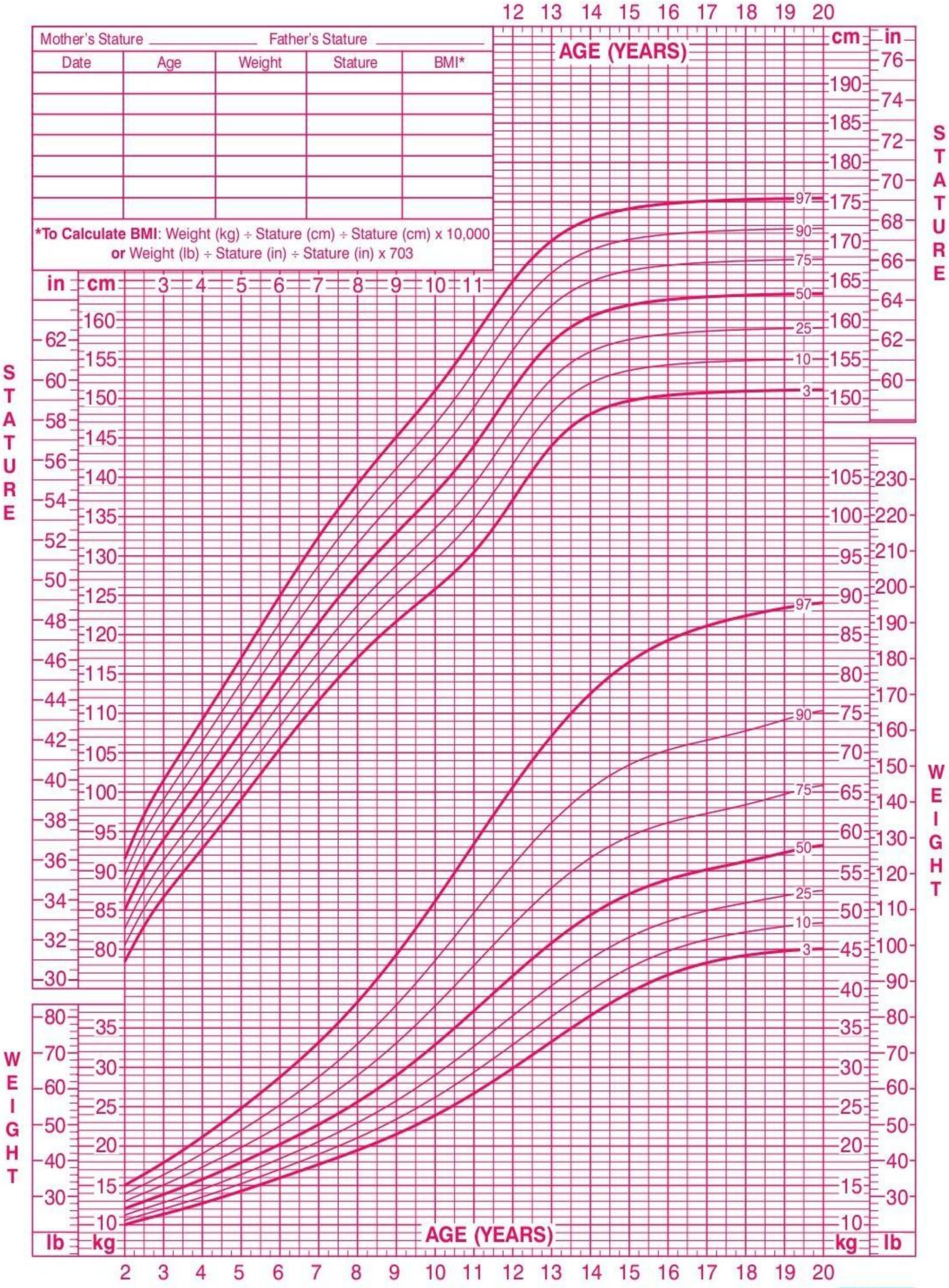


Published May 30, 2000 (modified 11/21/00).
SOURCE: Developed by the National Center for Health Statistics in collaboration with
the National Center for Chronic Disease Prevention and Health Promotion (2000).
http://www.cdc.gov/growthcharts

2 to 20 years: Girls
Stature-for-age and Weight-for-age percentiles

NAME ______________________

RECORD # ____________



Mother's Stature ____________ Father's Stature ____________

Date	Age	Weight	Stature	BMI*

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) x 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) x 703

Published May 30, 2000 (modified 11/21/00).
SOURCE: Developed by the National Center for Health Statistics in collaboration with
the National Center for Chronic Disease Prevention and Health Promotion (2000).
http://www.cdc.gov/growthcharts

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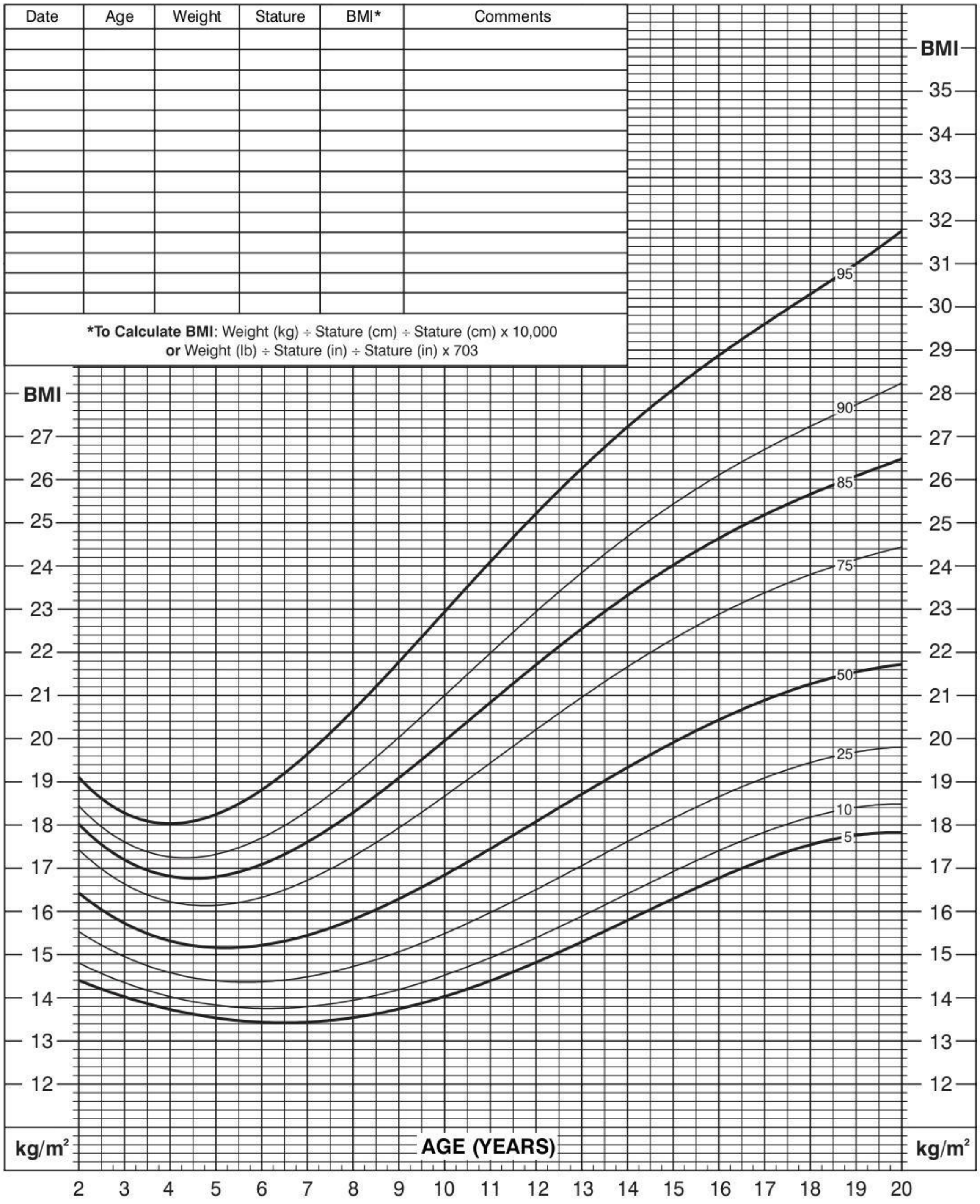
2 to 20 years: Girls
Body mass index-for-age percentiles

NAME ______________________

RECORD # ____________



Date	Age	Weight	Stature	BMI*	Comments

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) x 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) x 703

Published May 30, 2000 (modified 10/16/00).
SOURCE: Developed by the National Center for Health Statistics in collaboration with
the National Center for Chronic Disease Prevention and Health Promotion (2000).
http://www.cdc.gov/growthcharts

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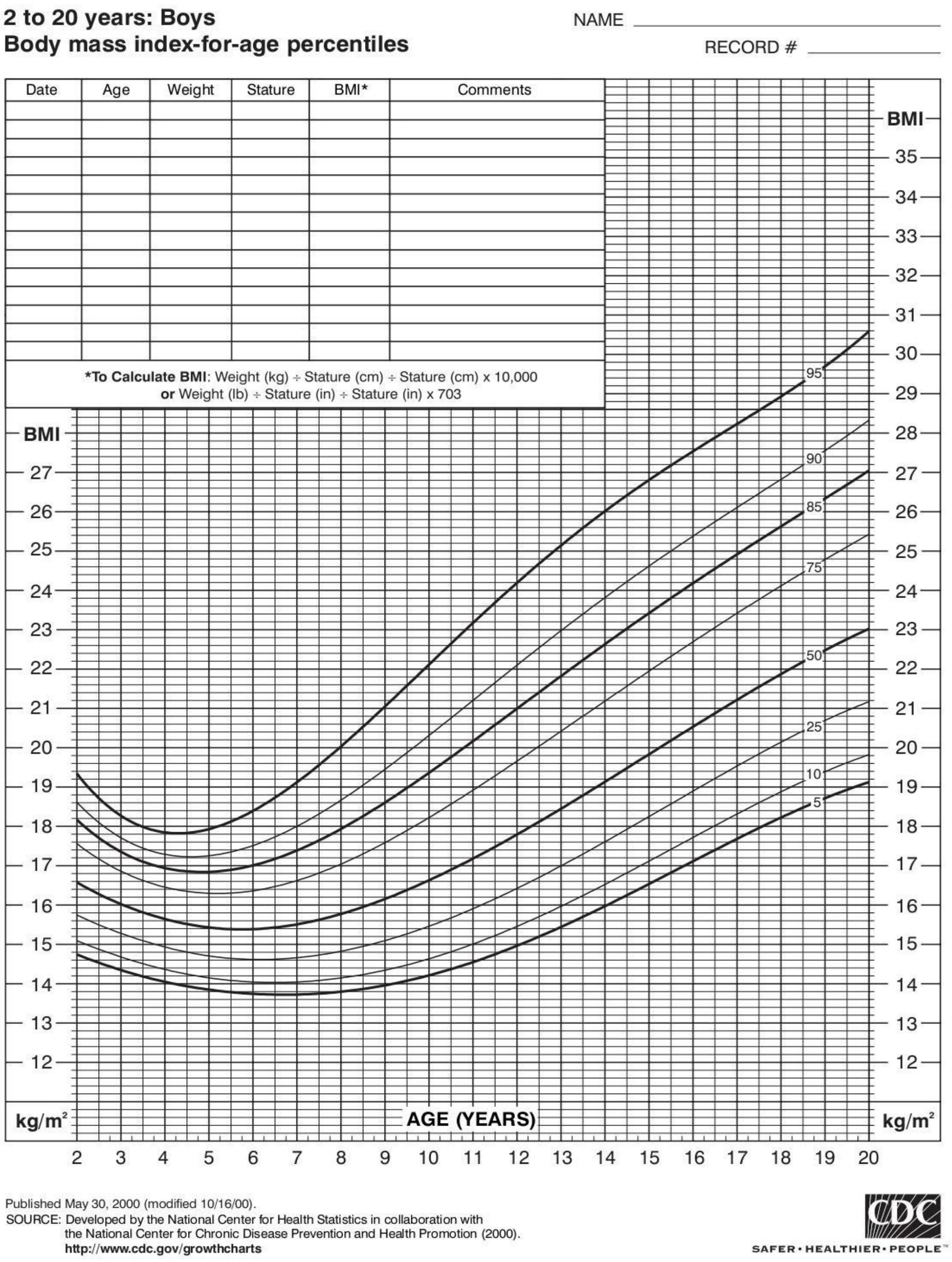
APPENDICIES



2 to 20 years: Boys
Body mass index-for-age percentiles

NAME ______________________

RECORD # ______________

Date	Age	Weight	Stature	BMI*	Comments

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) x 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) x 703

Published May 30, 2000 (modified 10/16/00).
SOURCE: Developed by the National Center for Health Statistics in collaboration with
the National Center for Chronic Disease Prevention and Health Promotion (2000).
http://www.cdc.gov/growthcharts

CDC
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APPENDIX (E)

Linguistic Approval

Ministry of Higher Education
and Scientific Research
University of Babylon
College of Education for Human Sciences

وزارة التعليم العالي والبحث العلمي
جمهورية العراق
جامعة بابل
كلية التربية للعلوم الانسانية

Ref. No
Date:

العدد : ٣٨١
التاريخ : ٢٠ /٦/ ٢٠٢٤

الواردة
العدد/ ١٦٥٢
التاريخ/ ٢٦/ ٦/ ٢٠٢٤

الى/ جامعة بابل / عمادة كلية التمريض / مكتب السيد معاون العميد للشؤون العلمية المحترم

م/ اعادة رسالة

تحية طيبة:

نعيد اليكم رسالة طالبة الماجستير (**نبأ حسين عيدان خدام**) في كليتكم بعد تقويمها لغويا من قبل (**أ.د.صالح مهدي عداي**) من قسم اللغة الانكليزية في كليتنا. نأمل من الباحثة الالتزام بالملاحظات المثبته على متن الرسالة .

..مع الاحترام..

أ.د. اسامة كاظم عمران
معاون العميد للشؤون العلمية
والدراسات والعليا

نسخة منه الى//
- الدراسات العليا .
- الصادرة

م/ العلمي . الرسائل

٢٦/٦

الخلاصة

اعتلال الهيموغلوبين هو مجموعة اضطرابات وراثية تنتقل من الاباء الى ابنائهم، ينتشر في جميع انحاء العالم، ويتميز بانخفاض أو غياب التوليف الطبيعي للهيموغلوبين كما هو الحال في الثلاسيميا، او بالتركيب غير الطبيعي للهيموغلوبين كما هو الحال في فقر الدم المنجلي، وله تأثير على الصحة الجسدية و النفسية الاجتماعية، حيث يؤدي الى زيادة المخاوف بشأن المظهر الجسدي والتدخل في عملية الحصول على الاستقلال والعلاقات الصحية مع العائلة والاصدقاء والمجتمع.

تهدف هذه الدراسة الى تقييم الصحة البدنية و النفسية الاجتماعية للمراهقين المصابين باضطرابات اعتلال الهيموغلوبين والكشف عن العلاقة بين الصحة البدنية والنفسية الاجتماعية مع بعض البيانات الديموغرافية وكذلك التحري عن العلاقة ما بين الصحة البدنية و النفسية الاجتماعية.

تم استخدام تصميم الدراسة الوصفية المقطعية خلال الفترة من 19 أكتوبر 2021 إلى 8 حزيران 2022 وقد أجريت هذه الدراسة في محافظة بابل على (150) مريضًا تتراوح أعمارهم بين 10-19 عامًا يعانون من اضطرابات الهيموجلوبين في مستشفى بابل التعليمي للنسائية والأطفال. و تم جمع البيانات باستخدام مقياس إلكتروني وشريط قياس غير قابل للمط واستبيان معدل صمم لغرض الدراسة، وقد تم تحليلها إلكترونيًا باستخدام برنامج الحزمة الاحصائية للعلوم الاجتماعية الاصدار 26.

أظهرت الدراسة أن الصحة البدنية العامة لأكثر من نصف العينة كانت معتدلة ، وأن الصحة النفسية والصحة الاجتماعية لأكثر من نصف العينة كانت جيدة. وقد ارتبطت بعض المتغيرات بالصحة البدنية ، مثل عمر المريض ، والمستوى التعليمي للمرضى وأولياء أمورهم ، ودرجة القرابة بين الوالدين ، ومستوى الهيموغلوبين. في حين أن الصحة النفسية والاجتماعية مرتبطة ببعض المتغيرات ، مثل المستوى التعليمي للمريض ، ووظيفة الأم ، والحالة الاقتصادية للأسرة ، ونوع المرض ، والمضاعفات. فضلا عن وجود علاقة معنوية بين الصحة البدنية والصحة النفسية و الاجتماعية بقيمة $p < 0.05$

خلصت الدراسة إلى وجود تأثير كبير لاضطرابات الهيموجلوبين على الصحة البدنية والنفسية والاجتماعية للمرضى المراهقين. وأوصت بتفعيل دور ممرضي الأطفال ومقدمي الرعاية الأسرية في تعزيز وتشجيع الأطفال المصابين باضطرابات اعتلال الهيموغلوبين لتحقيق أداء أفضل في أنشطتهم المعيشية اليومية ، وصحتهم النفسية ، والعلاقات الاجتماعية ، خاصة للأطفال الذين يعانون من قلة الدعم الاجتماعي وشدة المرض. لزيادة وعيهم من خلال الاهتمام بهم في جميع الجوانب التي تؤثر على شخصياتهم وتضعفها.

وزارة التعليم العالي والبحث العلمي

جامعة بابل

كلية التمريض

الصحة البدنية والنفسية الاجتماعية لدى المراهقين المصابين باضطرابات اعتلال الهيموغلوبين

رسالة تقدمت بها الطالبة

نبأ حسين عيدان

جامعة بابل/ كلية التمريض/ تمريض صحة الطفل

وهي جزء من متطلبات نيل درجة الماجستير علوم في التمريض

إشراف

أ. د. نهاد محمد قاسم الدوري

ذو القعدة 1443 هـ

حزيران 2022