Evaluation of Beta-Thalassemia effects on quality of life in school aged thalassemia patients

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Abstract :

Background : Thalassemia is one of the most common genetic disorders worldwide that associated with defective hemoglobin-chain synthesis. Clinical manifestations are diverse, ranging from asymptomatic hypochromia and microcytosis to profound anemia[1].

Research design : A cross sectional study was conducted using PedsQL questionnaire on 60 patient at Specialized Center for Hereditary Diseases at Babil Teaching Hospital for Maternity and Children from 22nd of February 2024 to the 14th of March 2024 .

The results of the research : showed that 33.3% of the sample have difficulty running , 46.7% complain of anger , 15% difficulty communication with others , 13.3% lack attention and 36.7% have to miss school for medical reasons.

As Conclusion Scheduled programs giving psychosocial help and a network connecting between the patients, school officials, thalassemia caregivers and the physician is required especially in developing countries where the health services are not integrated with social organizations , Special school services for thalassemia patients are required to deal with the repeated absence .

Recommendation : 1) for researcher's : Replication of the study on a large probability sample selected from different geographical areas in Iraq.

2)for patients : regular follow up for early detection of complication .

Aim of study :

1) Study the extent of thalassemia's effect on the patient's physical activity .

2) Study the extent of the effect of thalassemia on academic achievement .

3) Study the extent of the effect of thalassemia on the psychological aspect of the patient .

Introduction :

***Definition*:**

Thalassemia is an inherited blood condition. If you have it, your body has fewer red blood cells and less hemoglobin than it should. Hemoglobin is important because it lets your red blood cells carry oxygen to all parts of your body. Because of this, people with this condition may have anemia, which makes you feel tired.

You may hear it called things like Constant Spring, Cooley's anemia, or hemoglobin Bart's hydrops fetalis. These are common names for different forms of it. The two types are alpha thalassemia and beta thalassemia. The terms alpha and beta refer to the part of the hemoglobin the person is lacking.

There are also terms for how serious the thalassemia is. A person with a trait or minor form may not have symptoms or only mild ones. They may not need treatment. Someone with a major form will need medical treatment [2]

Causes:

Hemoglobin molecules are made of chains called alpha and beta chains that can be affected by mutations. In thalassemia there is inherited disorder caused by mutation in the globin genes that responsible of production either in β-chains or α-chain results in β-thalassemia or α-thalassemia respectively.

α-thalassemia: the severity of the disease is direct proportional to the number of α-globin genes that are deleted

β-thalassemia: the severity of thalassemia you have depends on which part of the hemoglobin molecule is affected[3]

Types:

1-Alpha-thalassemia:

There are four genes responsible for alpha-chain production, deletion involving one or more of the alpha-globin genes cause this type. As mentioned above the severity of the disease is direct proportional to the number of the deletion

- One gene deletion; Asymptomatic with no red cell abnormalities seen. The patient is silent carrier .
- Two genes deletion; May be asymptomatic or mild anemic, red cell abnormalities seen. Called alpha-thalassemia trait.
- Three genes deletion: Moderate to severe anemia. May not required blood transfusion. Cause *HbH Disease* "a relative excess of β-globin or (early in life) γ-globin chains. Excess β-globin and γ-globin chains form relatively stable β4 and γ4 tetramers known as HbH and Hb Bart, respectively. These types of hemogloin are functionally useless due abnormally high affinity to O2 that render the oxygen transport to the tissue."
- Four genes deletion: Rare but cause hydrop fetalis, babies born with this defect may die shortly after birth or require life long transfusion [4]

2-Beta-thalassemia:

It is the most common type of thalassemia, most prevalent in the Mediterranean area [4]. Two genes responsible of β-chains production . Mutations associated with β-thalassemia fall into two categories: (1) **β0**, in which no β-globin chains are produced; and (2**) β+**, in which there is reduced (but detectable) β-globin synthesis, causes three clinical syndrome depends of the type of mutation

Homozygous mutation: cause severe anemia and require regular blood transfusion, β-thalassemia major

- Variable mutation: Moderately severe anemia, regular blood transfusion not required, β-thalassemia Intermedia
- Heterozygous mutation: Asymptomatic with mild or absent anemia, red blood cell abnormalities seen, β-thalassemia minor [4]

- ***Diagnosis***

- Most children with moderate to severe thalassemia show signs and symptoms within their first two years of life. If your doctor suspects your child has thalassemia, he or she can confirm a diagnosis with blood tests.
- Blood tests can reveal the number of red blood cells and abnormalities in size, shape or color. Blood tests can also be used for DNA analysis to look for mutated genes.

- **Prenatal testing**

- Testing can be done before a baby is born to find out if he or she has thalassemia and determine how severe it might be. Tests used to diagnose thalassemia in fetuses include:

- **Chorionic villus sampling.** Usually done around the 11th week of pregnancy, this test involves removing a tiny piece of the placenta for evaluation.
- **Amniocentesis.** Usually done around the 16th week of pregnancy, this test involves examining a sample of the fluid that surrounds the fetus.[6]

- ***Complications***
- ***1-Skeletal Changes:***
- Red blood cell (RBC) production occurs primarily in the bone marrow. In the case of thalassemia, this RBC production is ineffective. One way the body attempts to improve production is by expanding the available space in the bone marrow. This most notably occurs in the bones of the skull and face. People can develop what is called "thalassemic facies"—chipmunk-like cheeks and a prominent forehead. Early initiation of chronic transfusion therapy can prevent this from occurring.
- Osteopenia (weak bones) and osteoporosis (thin and brittle bones) can occur in adolescents and young adults. It is not understood why these changes occur in thalassemia. The osteoporosis may be severe enough to cause fractures, particularly vertebral fractures. Transfusion therapy does not appear to prevent this complication.
- ***2-Splenomegaly***
- The spleen is capable of producing red blood cells (RBC); it generally loses this function around the fifth month of pregnancy. In thalassemia, the ineffective RBC production in the bone marrow can trigger the spleen to resume production. In an attempt to do this, the spleen grows in size (splenomegaly).
- This RBC production is not effective and does not improve anemia. Early initiation of transfusion therapy

can prevent this. If the splenomegaly causes an increase in transfusion volume and/or frequency, splenectomy (surgical removal of the spleen) may be required.

- ***3-Gallstones***
- Thalassemia is a hemolytic anemia, meaning the red blood cells are destroyed more rapidly than they can be produced. Destruction of the red blood cells releases bilirubin, a pigment, from the red blood cells. This excessive bilirubin may result in the development of multiple gallstones.
- In fact, more than half of people with beta thalassemia major will have gallstones by age 15. If the gallstones cause significant pain or inflammation, gallbladder removal (cholecystectomy) may be required.
- ***4-Iron Overload:***
- People with thalassemia are at risk for developing iron overload, also called hemochromatosis. Excessive iron comes from two sources: repeated red blood cell transfusions and/or increased absorption of iron from foods.
- Iron overload can cause significant medical problems in the heart, liver, and pancreas. Medications called iron chelators can be used to remove iron from the body.
- ***5-Aplastic Crisis***
- People with thalassemia (as well as other hemolytic anemias) require a high rate of new red blood cell production. Parvovirus B19 is a virus that causes a classic illness in children called Fifth Disease.
- Parvovirus infects the stem cells in the bone marrow, preventing RBC production for 7 to 10

days. This decrease in RBC production in a person with thalassemia leads to the development of severe anemia and usually the need for RBC transfusion.

- ***6-Endocrine Problems:***
- The excessive iron overload in thalassemia can result in iron being deposited in the endocrine organs, such as the pancreas, thyroid, and sex organs. Iron in the pancreas can result in the development of diabetes mellitus. Iron in the thyroid can cause hypothyroidism (low thyroid hormone levels), which may result in fatigue, weight gain, cold intolerance (feeling cold when others do not), and coarse hair. Iron in the sex organs may lead to symptoms that include decreased libido and impotence in men and lack of menstrual cycles in women.
- ***7-Heart and Lung Issues:***
- Heart issues are not uncommon in people with beta thalassemia major. Enlargement of the heart occurs early in life due to anemia. With less blood, the heart needs to pump harder, causing enlargement. Transfusion therapy can help prevent this from occurring. Long-term iron overload in the heart muscle is a major complication. Iron in the heart can cause an irregular heartbeat (arrhythmia) and heart failure. Starting iron chelation therapy early is crucial to preventing these life-threatening complications.
- Although the reasons why are not completely understood, people with thalassemia appear to be

at risk for developing pulmonary hypertension, or high blood pressure in the lungs. When blood pressure is elevated in the lungs, it makes it more difficult for the heart to pump blood into the lungs, which can lead to heart complications. Symptoms can be subtle, and screening tests are crucial so that treatment can be started early.[7]

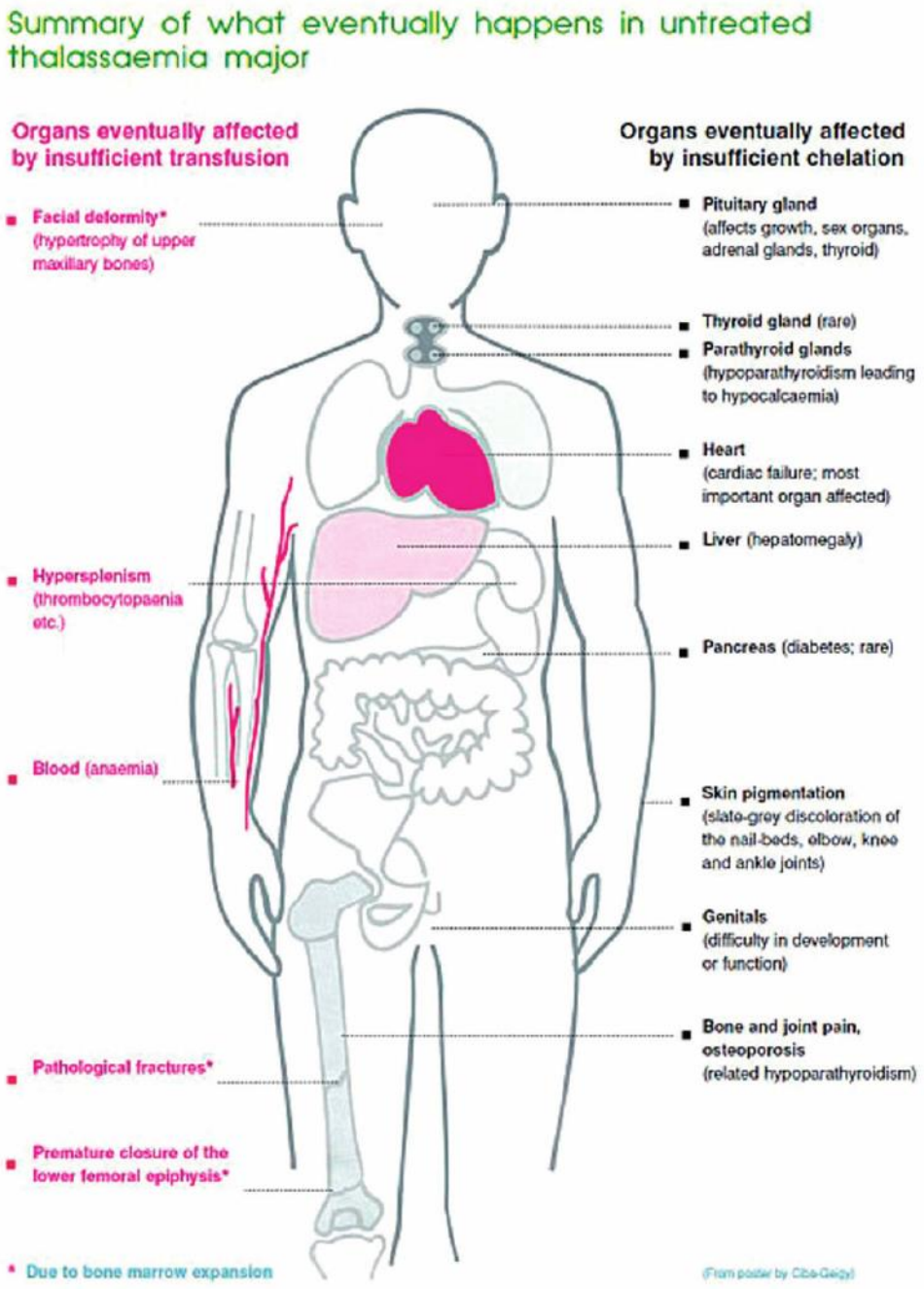

Figure 1. Complications of Thalassemia [14]

- ***Treatment:***
- Treatments for thalassemia depend on the type and how serious it is. If you are a carrier or have alpha or beta thalassemia trait, you likely have mild or no symptoms and may not need treatment.
- If you have a more serious thalassemia type like hemoglobin H disease, beta thalassemia intermedia, or beta thalassemia major you may experience moderate to serious anemia symptoms. You may need treatments such as blood transfusions, medicine, a splenectomy, or a blood and bone marrow transplant.
- -Blood transfusions
- Blood transfusions are the main way to treat moderate or severe thalassemia. This treatment gives you red blood cells with healthy hemoglobin.
- During a blood transfusion, a needle is used to insert an intravenous (IV) line into one of your blood vessels. You receive healthy blood through this line. The procedure usually takes 1 to 4 hours. How often blood transfusions are needed depends on how serious your condition and symptoms are.
- **Occasional blood transfusions** may be needed for people who have hemoglobin H disease or beta thalassemia intermedia. Specifically, a transfusion may be needed when your body is under stress, such as during an infection, pregnancy, or surgery.

- **Regular blood transfusions** (every 3 to 4 weeks) may be needed for people who have beta thalassemia major. These transfusions help maintain healthy hemoglobin and red blood cell levels.
- Iron chelation therapy
- The hemoglobin in red blood cells is an iron-rich protein . Regular blood transfusions can cause iron buildup, or iron overload, which can lead to potentially life-threatening complications.
- To prevent this, doctors use iron chelation therapy in people who receive regular blood transfusions to remove excess iron from the body. Three medicines are used for iron chelation therapy:
- **Deferasirox** is a pill taken once daily. Side effects can include skin rash, nausea, and diarrhea.
- **Deferiprone** is a pill that may be used if other treatments do not work. It can lower your white blood cell numbers, which can put you at risk for infections.
- **Deferoxamine** is a liquid medicine that is given slowly under the skin, usually with a small portable pump used overnight. This therapy takes time and can be mildly painful. Side effects can include problems with vision and hearing.
- Blood and bone marrow transplant
- A blood or bone marrow transplant, also called a hematopoietic stem cell transplant, replaces blood-forming stem cells that aren't working properly with healthy donor cells. A stem cell transplant is the only treatment that can cure thalassemia. However, only a small number of people who have severe thalassemia are able to find a good donor match and are a good fit for the procedure.
- Other treatments
- Even though blood transfusions are the typical treatment, other treatments may be used.

- **Medicines** called luspatercept (Reblozyl) and hydroxyurea may be prescribed by a healthcare provider to treat thalassemia. Luspatercept can lessen the number of blood transfusions needed for people with moderate to severe anemia as a result of thalassemia. Hydroxyurea is usually used to treat sickle cell disease and can help lower the risk of health problems from thalassemia.
- **Splenectomy** is surgery to remove the spleen. Your provider may recommend splenectomy to improve your symptoms if you have mild to moderate thalassemia. However, removing the spleen lowers the body's ability to fight infections.[8]

- ***Complication of Thalassemia Treatments***
- ***Iron Overload:***
- It occurs when iron intake is increased over a sustained period of time, either as a result of red blood cell transfusions or increased absorption of iron through the gastrointestinal (GI) tract. Both of these occur in thalassemia, with blood transfusion therapy being the major cause of iron overload in thalassemia major and increased GI absorption being more important in non-transfusion dependent thalassemia (NTDT). When thalassemia major patients receive regular blood transfusion, iron overload is inevitable because the human body lacks a mechanism to excrete excess iron. Iron accumulation is toxic to many tissues, causing heart failure, cirrhosis, liver cancer, growth retardation and multiple endocrine abnormalities[10]

Table 1, Distribution of complications related to iron overload in both TDT and NTDT [9]

	TDT	NTDT
Cardiovascular	Cardiac siderosis, left ventricular heart failure	Pulmonary hypertension, right ventricular heart failure, venous thrombosis
Liver	Liver fibrosis, liver cirrhosis, viral hepatitis	Liver fibrosis, liver cirrhosis, hepatocellular carcinoma
Endocrine	Hypothyroidism, hypoparathyroidism, growth retardation, hypogonadism, osteoporosis, diabetes mellitus	Osteoporosis
Other		Leg ulcers, gallstones, extramedullary hematopoietic tumors, silent cerebral ischemia

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- ***Blood Transfusion:***
- Gaining the most accurate information on the rate of iron loading from transfusion therapy is important in assisting selection of the best chelation therapy for each patient. A unit processed from 420 mL of donor blood contains approximately 200 mg of iron, or 0.47 mg/mL of whole donor blood. For red cell preparations with variable hematocrits, the iron mg/mL of blood can therefore be estimated from 1.16 × the hematocrit of the transfused blood product. In cases where organizational systems or other difficulties prevent such estimations to be calculated, a rough approximation can be made based on the assumption that 200 mg of iron is contained in each donor unit. Irrespective of whether the blood used is packed, semi-packed or diluted in additive solution, if the whole unit is given, this will approximate to 200 mg of iron intake. According to the recommended transfusion scheme for thalassemia major. the equivalent of 100–200 ml of pure red blood cell (RBC) per kg body weight per year are transfused. This is equivalent to 116-232 mg of iron/kg body weight / year, or 0.32-0.64 mg/kg/day.

Regular blood transfusion therapy therefore increases iron stores to many times the norm unless chelation treatment is provided[11]

- ***Increased gastro-intestinal absorption of iron:***
- In transfusion dependent thalassemia (TDT), the contribution of iron absorbed from the diet is small compared with blood transfusion. Normal intestinal iron absorption is about 1-2 mg/day. In patients with thalassemia who do not receive any transfusion, iron absorption increases several-fold. It has been estimated that iron absorption exceeds iron loss when expansion of red cell precursors in the bone marrow exceeds five times that of healthy individuals. Transfusion regimens aimed at keeping the pre-transfusion hemoglobin above 9 g/dl have been shown to prevent such expansion (Cazzola 1997). In individuals who are poorly transfused, absorption rises to 3-5 mg/day or more, representing an additional 1-2 g of iron loading per year[11]
- ***Iron Excretion:***
- The amount of iron in the human body is calculated through dietary consumption of iron, not through iron excretion. The removal of iron from the human body is handled at a very small rate. The fecal secretion of iron is equivalent to the consumption of iron by diet, and zero in the case of iron by transfusion. However, several cases such as blood transfusion in beta-thalassemia which suppress in production of hepcidin may lead to an overload of iron. Unintentional exposures can cause the majority of serious medical problems, such as cardiac arrhythmia and liver failure.

As a result, iron absorption is continuously monitored to prevent cell damage. The toxification of iron mainly occurs where it is stored i.e., in the liver. Biliary iron excretion estimated a significant amount of excess iron extraction through the bile and remaining to be processed for the reuse of hemoglobin synthesis. Urinary iron excretion a usual routine excretion of iron in the urine is a large proportion of the overall daily iron depletion[12]

- ***Diagnosis and quantification of iron overload:***
- 1. Magnetic resonance imaging;
- Magnetic resonance imaging (MRI) using R2 or T2* techniques has replaced liver biopsy as the gold standard for the quantification of LIC given its safety and reliability. Direct histological examination of hepatic tissue obtained by biopsy is highly sensitive and specific for the diagnosis of iron overload. As compared with MRI, liver biopsy remains more invasive, although it has low complication rates. Estimation of LIC by MRI
- in milligrams of iron per gram of liver dw reliably correlates with total body iron stores. MRI is also being used for the quantification of the cardiac iron concentration using T2* technique, in milliseconds. T2* gets shorter as myocardial iron concentration increases. Most guidelines for the management of thalassemia now rely on noninvasive monitoring using MRI to diagnose iron overload and tailor ICT[13]
- 2. Measure serum ferritin, iron, and transferrin saturation:
- Patients with ineffective erythropoiesis should be evaluated for secondary iron overload, which is diagnosed by measuring serum ferritin, serum iron,

and transferrin saturation. Serum ferritin measurement is the simplest and most direct initial test. Elevated levels (> 200 ng/mL [> 200 mcg/L] in women or > 250 ng/mL [> 250 mcg/L] in men) are usually present in secondary iron overload but can result from other abnormalities, such as hereditary hemochromatosis, inflammatory liver disorders (eg, chronic viral hepatitis, nonalcoholic fatty liver disease, alcoholic liver disease), cancer, certain systemic inflammatory disorders (eg, rheumatoid arthritis, hemophagocytic lymphohistiocytosis), or obesity.

- ***Therapies Under Investigation for Iron Overload:***
- 1. Hepcidin deficiency treatment:
- Hepcidin, the hormone which regulates the accumulation of absorption of iron and its abnormal transportation are a cause of iron overload in nearly every form of hereditary hemochromatosis and non-transfused iron overloading anemia.69 Analogues of hepcidin have also been seen reducing the toxicity of the iron-mediated tissue in mouse models.69 Agonists of hepcidin known as Mini-hepcidin are based on peptides that are rationally planned, based on the hepcidin field engaging with the ferroportin.60 Mini-hepcidin may be helpful in iron excessive conditions used for treatment or chelation treatment.70 Analogues of normal hepcidin and mini-hepcidin are studied for preventing the overloading of iron in hemochromatosis and beta-thalassemia. As deficiency in hepcidin causes an

overload of iron It would be expected that agents capable of mimicking hepcidin action or potentiating its endogenous production would inhibit iron.[12]

- 2-Gene therapy management:
- The treatment by gene therapy of genetic conditions such as sickle cell anemia and beta-thalassemia will prevent blood transfusions and reduce iron overload in the tissues. In individuals with hereditary hemochromatosis and beta-thalassemia including the DMT-1 activation and gene expression of ferroportin in enterocytes Over-expression of the wild-type HFE gene in enterocytes and overexpression of the iron regulatory peptide hepcidin in the liver are other therapeutic strategies that could be studied. The HFE genotype may influence the survival of myelodysplastic syndrome patients and tests need to be carried out if these patients are to be treated with effective iron chelation therapy.[12]
- ***Management of iron overload in thalassemia:***
- 1. Iron chelation therapy:
- an organ-based approach. The aims of ICT include maintenance of safe iron body stores to help counterbalance the excess iron loading, remove iron already deposited in tissues, and prompt reversal of heart failure. Three iron chelators have been approved by most regulatory agencies for ICT in thalassemia: DFO, DFP, and deferasirox (DFX).[13]
- 2. General guiding principles:
- Assessment of iron overload should be pursued after the transfusion of 10 U packed red blood cells in patients with TDT and at age of 10 years in patients with NTDT. Serum ferritin is used in early childhood

before patients can tolerate liver and myocardial MRI without sedation. Around age 8 to 10 years, consideration should be given to assessment of myocardial and hepatic iron load using noninvasive imaging techniques. delineates the proposed strategy for safety monitoring and follow up for ICT. The clinician should adopt an individualized approach when choosing ICT based on general iron overload profile, organ predominance of iron deposition, transfusion requirements, likelihood of adherence to therapy and follow-up, and profile of comorbidities.

- Oral ICT and novel formulations of previously approved agents may be better choices for patients with poor compliance. Our approach is to start DFO or DFX therapy for children with TDT older than 2 years of age. DFP serves as second line therapy, as experience with it has been the most limited. We also recommend close follow-up for early detection of no adherence or intolerance to therapy, which would warrant considering alternative agents and strategies. Long-term ICT was associated with better control of iron burden when therapy was appropriately and promptly adjusted for transfusional iron intake and patient weight based on set therapeutic goals.[14]

Materials and Methods :

Study population :A cross sectional study performed on 60 patients at Specialized Center for Hereditary Diseases at Babil Teaching Hospital for Maternity and Children , the samples were selected according to age from 22nd of February 2024 to the 14th of March 2024 (the visits were done on Saturday , Monday and Thursday between 9 Am. And 2 Pm.) . it included 30 males and 30 females and their age ranges between 7 years and 18 years old , they were all admitted to receive blood transfusion at the center .

Study instrument : in this study we used a modified Arabic version of PedsQL questionerre in order to assess quality of life in school age patients . The pedsQL include 22 items categorized into four main domains include : Physical health (seven items) , mental health (five items) , social health (five items) , school functioning (five items) Also patient name , sex , age , number of blood transfusion per month and usage of iron chelating factors were documented.

Data collection : children and/or their parents were interviewed using Pediatric Quality of Life inventory 4.0 Generic Core Scale , giving clear explanation and enough time to every item by the medical students who did the interview .

Data analysis : The collected data were analyzed manually . Patient characteristics and demographics information were showed by percentage , pie charts and bar graph

Pilot study :

A pilot study was carried out on 10% (6) of the patients under study to evaluate the applicability, clarity, efficiency and time needed to fill in each tool. Also, it help to find the possible obstacles and problems that might face the researcher and interfere with data collection. The necessary modifications were done, where some items and questions were omitted and others were added based on the result of the pilot study. The six patients who included in the pilot study were excluded from the study.

1) Age of patients :

Summary statistic	***Value***
Mean	13,83
Median	14.5
Mode	17
Standard deviation	3,37
Minimum	9
Maximum	17

2) Blood transfusion frequency :

	One per month	***Two per month***	***more***
Frequency	50%	16,7%	33,3%

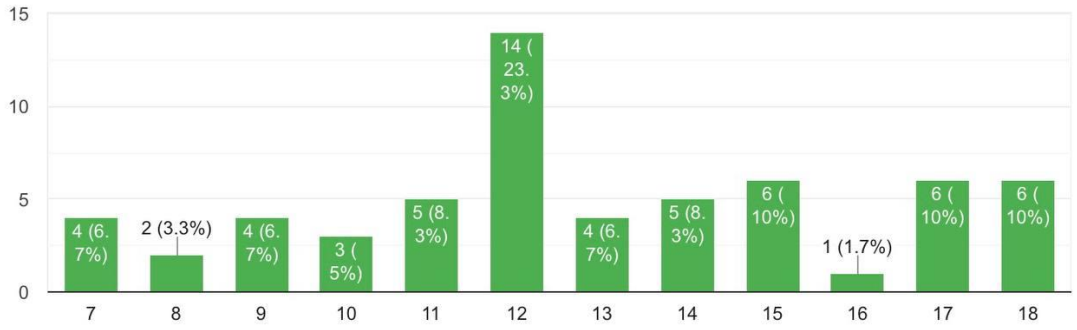
3) Quality of life :

	Yes	***Sometimes***	***No***
Difficulty in running	16,7%	33,3%	50%
Physical pain	16,7%	66,7%	16,7%
Feeling angry	66,6%	16,7%	16,7%
Difficult communicating with others	0%	16,7%	83,3%
Difficulty in attention and focus	33,3%	33,3%	33,3%
Absence from school	100%	0%	0%

Results :

1) Age of patients :

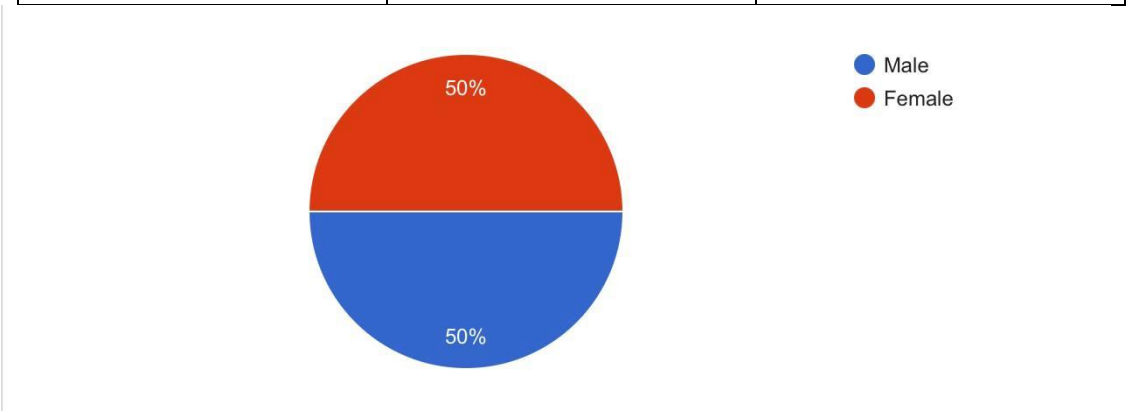
Summary statistic	***Value***
Mean	12,85
Median	12
Mode	12
Standard deviation	3,193
Minimum	7
Maximum	18



Bar graph 1 : age

2) <u>Gender</u> :

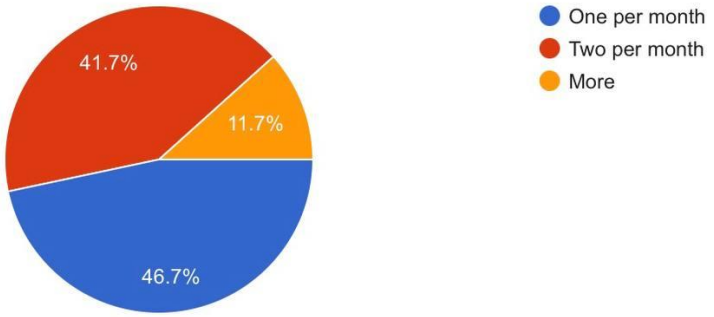
	Number	Percentage
Male	30	50%
Female	30	50%



Pie chart 1: gender of patient.

3) <u>Blood transfusion frequency</u> :

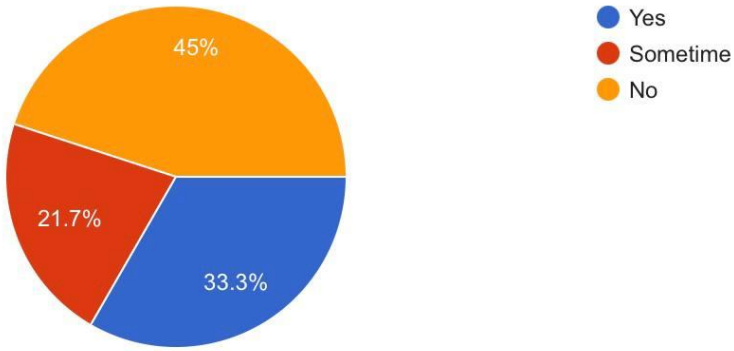
Frequency	Number of patient	Percentage
One per month	28	46,7%
Two per month	25	41,7%
more	7	11,7%



Pie chart 2 : blood transfusion frequency.

4) <u>difficulty in running</u> :

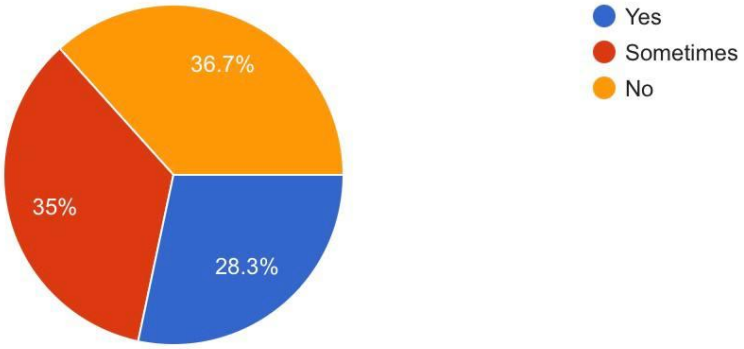
	Number of patient	Percentage
Yes	20	33,3%
Sometimes	13	21,7%
No	27	45%



Pie chart 3: difficulty in running.

5) <u>Physical pain</u> :

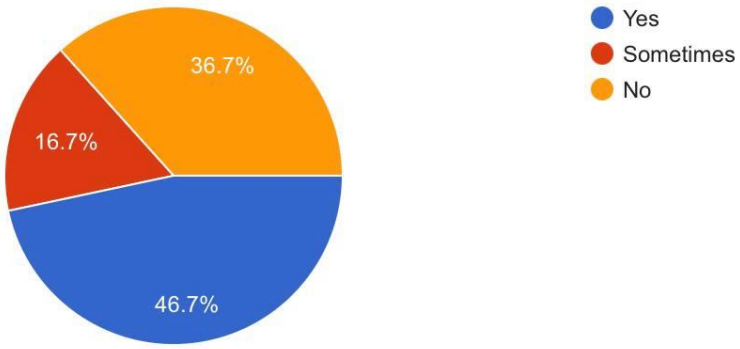
	Number of patient	Percentage
Yes	17	28,3%
Sometimes	21	35%
No	22	36,7%



Pie chart 4 : physical pain.

6) <u>Feeling of anger</u> :

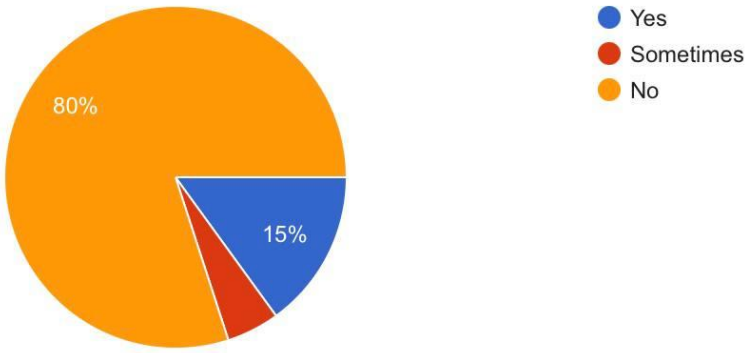
	Number of patient	Percentage
Yes	28	46,7%
Sometimes	10	16,7%
No	22	36,7%



Pie chart 5: feeling of anger .

7) <u>Difficulty communication with others</u> :

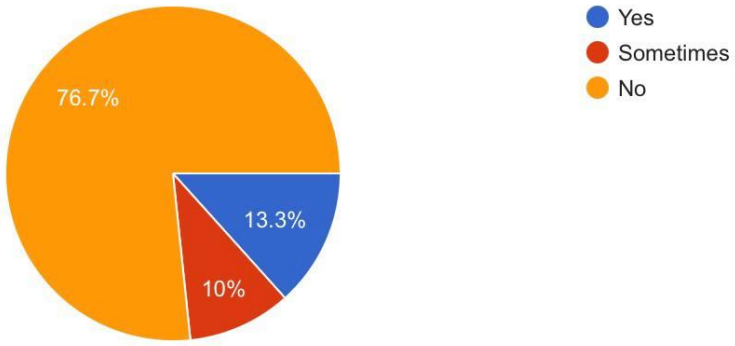
	Number of patient	Percentage
Yes	9	15%
Sometimes	3	5%
No	48	80%



Pie chart 6 : difficulty in communication with others

8) Difficulty in attention and focus :

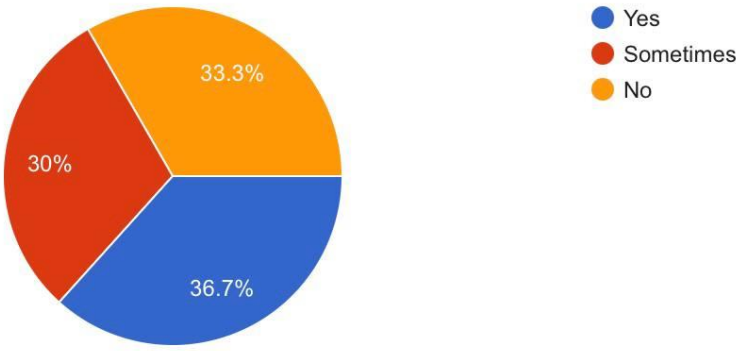
	Number of patient	Percentage
Yes	8	13,3%
Sometimes	6	10%
No	46	76,7%



Pie chart 7 : difficulty in attention and focus .

9) Absence from school for their medical condition :

	Number of patient	Percentage
Yes	22	36,7%
Sometimes	18	30%
No	20	33,3%



Pie chart 8 : absence from school for their medical condtion .

Discussion :

Thalassemia is the most common form of inherited anemia worldwide [15].The chronic nature of the disease and complications associated with clinical signs of the disease and its treatment make multiple physical, psychological and social

problems and effects on the quality of life in these patients. Therefore, assessing knowledge and QoL of these patients leads to better understanding of their specific needs and using more effective care [16].

Concerning the gender , the study illustrated that there is equally percentage for male and female gender.

This results disagreed with Taheri etc.[17] who revealed that more than half of the study subjects were males and more than one third of subjects were females in a study .

And this difference maybe due to small sample size in our study.

Concerning the activity of thalassemia patient (difficulty in running), the present study illustrated that (33,3%) of them suffering from difficulty in running, (21,7%) sometime and (45%) with no difficulty.

The present results agree with Kharyal et al., (2021)[18] who revealed that less than half of patients know that physical activities of thalassemia patients are affected by the level of hemoglobin and dyspnea aggravated by daily activity in a study that titled "Disease knowledge and general self-efficacy among adolescents with thalassemia major and their parents' perspective.

Concerning the physical pain of thalassemia patient , the present study illustrated that (28,3%) with pain, (35%) sometimes and (36,7%) with no pain .

This results agreed with Sinlapamongkolkul Phakatip etc.[19] who revealed that the most problematic was having pain or discomfort .

Concerning the feeling angry in thalassemia patient , the data show high percentage feeling angry (46,7%) ,(16,7%) sometimes while just (36,7%) with no feeling angry.

This results agreed with Ghanizadeh Ahmed etc.[20] who revealed that (62,7%)suffered from irritability and anger.

Concerning the difficulty in communication with others , there is (80%) with no difficulty or problem , while just (15%) with difficulty and (5%) sometimes .

This results disagreed with Messina Giuseppina etc.[21] who revealed that the emotional role and social function values were considerable lower than in all of the domain . while our study show the patients are normal in communication with others.

And this difference might be duo to high prevalence of thalassemia in Iraq.

Concerning the difficulty in attention and focus , the data show there is (76,7%) with no difficulty , (13,3%) with difficulty and (10%) with sometimes .

This results disagreed with Uni Gamayani etc.[22] who revealed that thalassemia patient have significantly lower attention , verbal memory capability and executive function.

This difference can be explained by the dependence of our study on patient's responses , while the referred research used a specific tools and tests to calculate cognitive functioning .

Concerning the absence from school , the study illustrated that there is (36,7%) absence from school for there medical condition and (30%) with sometimes absence while (33,3%) with no absence from school .

This results agreed with Dina K. Ismail etc.[23] who revealed that there is a significant decrease in school attendance of thalassemia group compared with that control group.

Conclusion :

- about half the sample suffer from irritability and anger problems.
- most of the patients has no trouble getting along with other

kids or making friends.

- duo to closure of the transfusion center on holidays , patients has to miss school in order to get treatment.
- relating to physical health , patients complained of fatigue and tiredness in the days prior to transfusion.
- Special school services for thalassemia patients are required to deal with the repeated absence .
- Scheduled programs giving psychosocial help and a network connecting between the patients, school officials, thalassemia caregivers and the physician is required

Questionnaire :

العمر:

☐ ذكر ☐ انثى

عدد مرات نقل الدم شهريا :

: Iron chelating agent intake

الصحة الجسدية	نعم	احيانا	لا
اواجه مشكلة في المشي لمسافة قليلة			
اواجه صعوبة في الركض			
اواجه صعوبة في المشاركة برياضة معينه			
لا استطيع حمل أشياء ثقيلة			
اواجه صعوبة في المساعدة بالأعمال المنزلية			

اعاني من الام جسدية			
اعشر بالتعب اغلب الأوقات			

الصحة النفسية	نعم	احيانا	لا
اشعر بالخوف			
اشعر بالحزن			
اشعر بالغضب			
اشعر بصعوبة في النوم			
اشعر بالقلق من المستقبل			

الصحة الاجتماعية	نعم	احيانا	لا
اواجه صعوبة في الاندماج مع الاخرين			
الناس لا يرغبون في تكوين صداقات معي			
اتعرض للمضايقات من الاخرين			
لا استطيع القيام بما يقوم بها الاخرين من نفس الفئة			
لا استطيع الاستمرار باللعب مع الاخرين			

الوظيفة/المدرسة	نعم	أحيانا	لا
اواجه مشكلة في الانتباه			
اعاني من كثرة النسيان			
لا استطيع مواكبة الواجبات			
الغياب عن المدرسة بسبب الحالة الصحية			
الغياب عن المدرسة بسبب زيارة الطبيب			

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