Compliance of Children with Beta Major Thalassemia to their Receiving of Iron Chelation Therapy

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Abstract

Background: Beta major thalassemia is a hereditary hemoglobin disorder that results from absence of a beta-globin chain in the pathway of hemoglobin production. It requires regular blood transfusion which leads to iron overload that requires iron chelation therapy. **Aim of the study:** Was to assess the compliance of children with beta major thalassemia to their receiving of iron chelation therapy. **Study design:** A descriptive research design was utilized to conduct this study. **Setting:** The study was conducted at medical building at fourth floor in hematological diseases unit at Banha Specialized Pediatric Hospital affiliated to secretariat of specialized medical center. **Subjects:** A purposive sample of 100 children diagnosed with beta major thalassemia and their mothers who attended the previously mentioned setting. **Tools of data collection:** Two tools were used; **Tool I:** A structured Interviewing Questionnaire Sheet. **Tool II:** Reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene. **Result:** Vast majority of the studied children are not complying to iron chelation therapy. Also, majority of the studied mother had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. And also, more than three quarters of the studied children had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. Furthermore, more than half of the studied children had satisfactory practices regarding total reported practice regarding steps of giving Oxjade. and more than half of the studied mothers have in satisfactory practices regarding total reported practice regarding steps of administering disferal medication. **Conclusion:** More than half of the studied children are not compliance to iron chelation therapy. While, less than half of them are compliance to iron chelation therapy. **Recommendations:** Emphasize the role of nurses in education the thalassemic children and their mothers regarding compliance to iron chelation therapy.

Keywords: Compliance, Children, Beta major thalassemia and Iron chelation therapy.

Introduction:

Beta major thalassemia or Cooley anemia, is a prevalent monogenic genetic hemoglobin disease. It arises from the hemoglobin synthesis pathway's lack of a beta-globin chain. More than 200 beta-globin gene point mutations cause beta-thalassemia, a genetic condition characterized by reduced or missing globin chains (beta+ and beta0). Ineffective erythropoiesis is the main underlying pathology. Children with major thalassemia consequently develop a lifelong reliance on blood transfusions. This illness needs to be identified and treated right away to prevent the significant morbidity and mortality rates linked to it **(Khan & Shaikh, 2023).** Globally, thalassemia is the most common monogenic disease. It is a genetic condition marked by quantitative abnormalities in globin. From 1.5 to 7% of the global population is thought to be carriers of thalassemia genetic mutation. Southeast Asia, the Middle East, the Indian subcontinent, the Mediterranean region, and portions of North and sub-Saharan Africa are

where thalassemia is most common. Due to continuous migration, these illnesses are becoming more prevalent in major multiethnic cities in North America and Europe, which is a global health concern **(Li et al., 2024).**

Children with major beta thalassemia exhibit hepatosplenomegaly, growth failure, recurrent infections, abdominal distension, anemia, and progressive pallor. Hemolytic features include frontal bossing, maxillary prominence, hypertelorism, epicanthic folds, and dental malocclusion, as well as skin discoloration brought on by hemosiderosis and pallor. Complete blood counts can be used to diagnose children with thalassemia: increased reticulocyte count, low MCV and MCH, and a hemoglobin level ranging from 2 to 8 g/dL. Red cell morphological anomalies such as hypochromia, microcytosis, and polychromatophilic cells are seen in peripheral smears. Analysis of Hb, both qualitative and quantitative (using cellulose acetate electrophoresis): determines the kind and quantity of Hb that is present. Radiology: Hand and skull X-rays. Elevated serum bilirubin **(Pal, 2021).**

Children with thalassemia need frequent blood transfusions to boost their hemoglobin levels and treat their anemia. Serum ferritin levels will be impacted by repeated blood transfusion sessions to accomplish these objectives. Iron buildup in the heart, liver, spleen, and other organs due to systemic iron excess can result in a several problems. Iron chelation therapy (ICT) is necessary to effectively address the concerning clinical problem of the potential of severe iron excess consequences. Three iron chelators are now on the market: deferoxamine (DFO), deferiprone (DFP), and deferasirox (DFX). ICT is essential for managing post-transfusional iron overload **(Lee et al., 2024).**

Compliance is known as the degree to which a child's conduct (taking medicine, following a diet, and altering their lifestyle) complies with a healthcare provider's instructions. Adherence is a complicated phenomenon that involves psychological elements as well as interconnected elements linked to children's health, condition, treatment, and surroundings. These elements all affect how effectively a child follows a recommended routine. Methods of measuring compliance can be divided into two categories: direct methods and indirect approaches. Drug concentration measurements in blood and directly observed therapy are examples of direct approaches. Child’s self-report, pill counts, clinical response evaluation, and the child and mother report or questionnaire are examples of indirect approaches **(Mohamed et al., 2022).**

Deferoxamine was identified as a chelation agent that used intramuscularly. It worked best when administered subcutaneously over a period of 8 to 24 hours. Pain and swelling at the infusion site are common side effects of subcutaneous infusions, that involve inserting a needle into subcutaneous tissue on the legs or abdomen. Deferasirox This is taken orally once a day and can be used either alone or in conjunction with the other medication. Abdominal pain, diarrhea, vomiting, and nausea are typical adverse effects. Deferiprone is available in tablet and syrup form and is taken three times a day. Typical adverse effects include arthralgias, gastrointestinal side effects and a low white cell count **(Eziefula et al., 2022).**

Nurses play a critical role in devising effective educational programs for children and their caregivers. Spreading information about the disease's pathophysiology, hereditary transmission, the importance of treatment adherence, the use of a subcutaneous pump at

home and oxajad to administer iron chelation therapy as directed, possible side effects, and more. Additionally, teaching patients and caregivers about anemia, its symptoms, effects, management strategies, the need for blood transfusions, and managing its related complications. Furthermore, since βthalassemia can result in increased gastrointestinal absorption of iron and subsequent iron overload, it is imperative to educate people about dietary considerations. Iron-rich foods such as liver, leafy greens, grains, spleen, molasses, legumes, and nuts should be avoided by children **(Mahfoz et al., 2024).**

Significance of the study:

Beta major thalassemia is the most common and severe form of thalassemia that known as thalassemia major or Cooley's anemia. Beta major thalassemia is a heterogeneous group of hereditary hemoglobinopathies characterized by defects in the β-globin chain of hemoglobin and belong to autosomal recessive disorders, it affects mainly persons in the Mediterranean region, but they also occur in populations living in Middle East, Transcaucasia, Central Asia, Indian subcontinent, Far East, and Africa **(Kattamis et al., 2022).** World Health Organization estimated an incidence of β-thalassemia of 40,000 newborns each year and a total annual incidence of symptomatic children about 1/100,000 people around the world and 1/10,000 people in Europe **(Russo et al., 2016).** In the United States, it was reported that 1: 272, 000 live births **(United States of America, 2020).** It has been estimated that one thousand children out of 1.5 million live births are born each year suffering from thalassemia in Egypt. It is reported that the carrier rate in Egypt is between 9 to 10% of the population **(El-Shanshory et al., 2021).** So the researchers will carry out this study to assess compliance of children with beta major thalassemia to iron chelation therapy.

Aim of the Study:

The aim of this study was to assess the compliance of children with beta major thalassemia to their receiving of iron chelation therapy.

Research Hypotheses:

To what extent do children with beta major thalassemia comply to their receiving of iron chelation therapy?

Subjects & Method:

Research Design:

A descriptive research design was utilized to conduct this study.

Study Setting:

The study was conducted at medical building at fourth floor in hematological diseases unit at Banha Specialized Pediatric Hospital affiliated to secretariat of specialized medical center. It had 2 rooms (the first room had 21bed, the second room is an isolating room and had one bed).

Study Subjects:

The sample size was calculated using this A purposive sample of 100 children diagnosed with beta major thalassemia and their mothers who attended the previously mentioned setting, the children were comprised the following criteria, Age ranges from 6-15 years, free from any other chronic diseases and receiving of iron chelation therapy. The study was carried out through a period of three months.

Tools of data collection: Data was collected through the following tool:

Tool I: A structured Interviewing Questionnaire Sheet: It was adopted by **Wahyuni et al., 2011; Elsoudy et al., 2022.** It was used to assess compliance of children with beta major thalassemia to their iron chelation therapy. It was written in a simple Arabic language to suit the children's and mother's level of understanding. It was composed of 5 parts as following:

Part 1:- Characteristics of Children with Beta Major Thalassemia: This part was concerned with personal characteristics of children with beta major thalassemia. It includes 5 items regarding age in years, gender, educational level, child's rank and residence.

Part 2:- Medical History of Children: This part was concerned with medical history of children. It included 15 items, including: time when the disease start appearing, tests that is done to diagnose child condition, routine test that the child do, child receiving of blood transfusion, number of times the blood transfused, if child have ever been enter the hospital, if enter what is the reason, If the reason is related to the disease, what is it, child compliance to iron chelation therapy, type of iron chelating medication that the child takes, child have any complication after receiving the medication, what this complication, number of times the complication occurred after receiving iron chelation therapy, child have any problems while receiving blood cells and what these problems.

Part 3:- Compliance of Children with Beta Major Thalassemia to their Iron Chelation Therapy: This part was concerned with assessing compliance of children with beta major thalassemia to their Iron chelation therapy. It was adopted from **Wahyuni et al., 2011; Elsoudy et al., 2022.** It included five main items, first item: The degree of child commitment to the proper nutrition which include 4 sub items as; reducing iron rich foods such as eggplant and liver, eat food that help absorption of iron, such as orange, lemon and green pepper, reducing foods that contain high percentage of preservative, such as chips and pepsi, eat large amounts of foods that contain folic acid, including lettuce and okra. Second item: The degree of child commitment to medication which include 4 sub items as; take the right prescribed dose, take vaccinations regularly to prevent any infection, receiving preventive antibiotics according to doctor order. Third item: compliance of children with thalassemia with blood transfusion which include 4 sub items as; visiting hospital regularly for blood transfusion, following doctor order regarding daily activity after blood transfusion, following doctor order regarding diet, inform doctor immediately if any health problem occurs after blood transfusion. Fourth item: Compliance of Children with Thalassemia to Personal Hygiene: which include 7 sub items as; washing hands regularly, cutting nails regularly, taking care of hair, taking bath every day, cleaning teeth regularly, using the child of his own tools, not sharing them with one of his siblings. Fifth item: Compliance of children with Thalassemia to Follow Up: which include 6 sub items as; visiting a doctor regularly for check-up, following the doctor's instructions according to diet, measuring the weight regularly, doing a periodic check-up on teeth regularly, informing doctors about child's condition before any medical tests or surgical operation, visiting the doctor immediately when any deterioration in health occurs.

Scoring system for children's compliance:

Each item would be rated on a three points Likert scale as the following: always complying took score (2), sometimes complying took score (1), never complying took score (0). **The total scoring system for child's compliance was calculated and classified as the following:**

-Totally compliance took score (75%-100%)
-Sometimes compliance took score (60%-< 75%)
- Never compliance took score (< 60 %-50%).

Part 4: -Mother's and children's knowledge regarding beta thalassemia and blood component:

This part was concerned with mother's and children's knowledge regarding beta thalassemia and blood component. It adapted

from **Berman et al., 2008.** It included 14 questions, including: components of blood, main place for the manufacture of blood and blood products, function of red blood cells, definition of thalassemia, the causes of thalassemia, symptoms of thalassemia, complications of thalassemia, result of noncompliance to treatment , basic treatment for a child suffering from thalassemia, complications of blood transfusion, methods of treating blood transfusion complications, ways to prevent thalassemia, importance of treatment with Disferal or Oxjade, side effects of Disferal, foods that is rich in iron.

Scoring system for child's and mother's knowledge:

The studied sample knowledge was checked with a model key answer, as the following scores (1) were given for correct answer and score (0) for incorrect answer. **The total scoring system for child's and mother's knowledge was calculated and classified as the following:**

-Satisfactory reported practice was taken score of (≥75%).

- Unsatisfactory reported practice was taken score of (<75%).

Tool II: Mother's and children's reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene: This part was concerned mother's and children's reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene. It adapted from **Berman et al., 2008.** It included four main items First: the procedural steps for washing hands and it included 10 steps, Second - the procedural steps for administering disferal medication and it included 14 steps, third: the procedural steps for oral care and it include 5 steps, fourth: the procedural steps for giving oxajade medication and it included 8 steps.

Scoring system for mother's and children's reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene:

Score (1) was given for steps that done correctly; score (0) was given for steps that done incorrectly.

The total scoring system for mother's and children's reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene was calculated and classified as the following:

-Satisfactory reported practice was taken score of (≥75%).

- Unsatisfactory reported practice was taken score of (<75%).

Content validity:

Tools for data collection were revised by a panel of three experts in the field of Pediatric Nursing Department, at Faculty of Nursing Benha University. They reviewed the tools for its clarity, relevance, comprehensiveness, simplicity and applicability and minor modifications were done according to their judgment. This modification in tools as the following (adding a part about mother's and children's reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene, so the pilot sample was excluded from the study

Reliability of the tools:

Reliability of the developed tools was estimated by using Cronbach alpha test to measure the internal consistency of the study tools, Results from repeated testing was compared (test- retest reliability). Where it was found that: Children's compliance r = 0.964, Children's knowledge r = 0.809, Children's reported practice r = 0.823, Mother's knowledge r =0.797, Mother's reported practice r =0.879. These results indicate high degree of reliability for the study tools.

Ethical considerations:

The researchers took oral consent from the mother of children before their participation in the study and approval of scientific ethical

research committee. Each child was secured that the study not carried any physical, psychological and social risks. They informed that all the gathered data treated confidentially and used for the research purpose only and they have the right to withdraw at any time from the study.

Pilot study:

A pilot study was carried out on 10% from children (10 children and their mother) of the expected total sample size to test the feasibility, clarity, objectivity, applicability and time needed for data collection. It was done at a period of one month (beginning of April 2024). There is no vital modification in tool so the pilot study was included in the study.

Field work:

The data will be collected from the previously mentioned setting according to the policy of the setting. The researchers started by introducing herself to the mothers and their children and explaining the aim of the study and took their oral approval to participate in the study prior to data collection. The researchers used the previously mentioned study tools to assess compliance of children to iron chelation therapy. Data collection took a period of three months starting from the beginning of May 2024 up to the end of July 2024, 2 days per week (Sunday & Wednesday), 3 hours per day from (10 am to 1 pm) in the previously mentioned setting in the morning shift according to hospital policy. The researchers took (4-5) children and their mother per day. the researchers collect data from them by the previously mentioned tools. The researchers interviewed individually with each mother and her child and took a period of 1 hour with each mother to complete the questionnaire as the following (5 min for personal characteristic of mother and child , 10 min for medical history of child , 10-15 min for compliance of child to iron chelation therapy , 10 min for mother's and child's knowledge regarding beta thalassemia and blood component ,20 min for mother's and child's reported practice regarding steps of giving disferal and oxjade medication, steps of mouth care and hand hygiene)

Statistical analysis:

The data obtained were organized, categorized, analyzed, and presented in the form of tables and figures using the statistical package for socialist in version 20 (SPS), Qualitative variables were presented in the form of frequencies and percentages, quantitative variables were presented in the form mean and Standard Deviation. Test of significance were used to find out associations between study variables. Chi-square (x2) test of significance was used in order to compare proportions between two qualitative parameters. Spearman's rank correlation coefficient (r) was used to assess the correlation between two variables. The confidence interval was set to 95% and the margin of error accepted was set to 5%.

Results:

Table (1): Shows that, less than half (46%) of the studied children were in the age group 12-<15 years, with mean age of 12.71±1.71 years. Concerning gender, less than two thirds (64.0%) of them are male. Also, more than half (54.0%) of them were in the primary education level. Moreover, less than two thirds (60.0%) of them were the first child between their siblings. In addition, more than half (59.0%) of them residing in urban area.

Table (2): Presents that, the majority (83.0%) of the studied children have a disease from infanthood. Also, all (100.0%) of them have a hemoglobin electrophoresis test to diagnose the disease and have a complete blood count as a routine. Moreover, the vast majority (95.0%) of the studied children receive blood transfusion, (75.8%) of them receive once every three weeks. In addition, the majority (80.0%) of the studied children have history

from hospitalization, (96.3%) of them hospitalized due to disease and 96.0% of them hospitalized to receive treatment of the disease. Also, the vast majority (94.0%) of them are adherent with iron chelation therapy. Moreover, less than three quarters (74.0%) of them take Oxajade. Furthermore, the minority (16.0%) of the studied children have history from complication after receiving the medication, all (100.0%) of them have vomiting and 81.3% of them have complications that occurred twice after receiving iron chelation therapy .

Fig (1): Shows that, more than half (58.0%) of the studied children are not compliance to iron chelation therapy. While, less than half (42.0%) of them are compliance to iron chelation therapy

Fig (2): Shows that, more than half (53.0%) of the studied mother had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. While, less than half (47.0%) of them had satisfactory level of total knowledge. It also shows that, more than three quarters (76.0%) of the studied children had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. While, nearly less than one quarter (24.0%) of them have satisfactory level of total knowledge.

Fig (3): Illustrates that, more than half (54.1%) of the studied children had unsatisfactory practices regarding total reported practice regarding steps of giving Oxjade. While, less than half (45.9%) of them had satisfactory practices.

Fig (4): Displays that, more than half (53.8%) of the studied mothers have unsatisfactory practices regarding total reported practice regarding steps of administering disferal medication. While, less than half (46.2%) of them have satisfactory practices.

Table (3): Demonstrates that, there was highly statistically significant relation between total children's compliance with iron chelation therapy and their age and educational level at P= < 0.01. While, there was no statistically significant relation with their gender, ranking and residence at P= > 0.05

Table (4): Clarifies that, there are highly statistically significant relation between total children's compliance with iron chelation therapy and their family history from beta major thalassemia P= < 0.01. While, there are no statistically significant relation with their age at onset of disease, receive blood transfusion, history from hospitalization, type of iron chelating medication and history from complication after receiving the medication at (P= > 0.05 .(

Table (5): Indicates that, there are highly significant positive correlation between total children's compliance score and their total knowledge score (r=0.717, p=<0.01), and total reported practices score (r=0.903, p=<0.01). Also, there are highly significant positive correlation between total children's knowledge score and their total reported practices score (r=0.610, p=<0.01

Table (6): Indicates that, there are highly significant positive correlation between total mother's knowledge score and their total reported practices score (r=0.910, p=<0.01.

Table (1): Distribution of the studied children according to their personal characteristics (n=100).

Personal characteristics	No.	%
Age (years)		
6- <9	11	11.0
9- <12	43	43.0
12- <15	46	**46.0**
Mean ± SD	**12.71±1.71**	
Gender		
Male	64	**64.0**
Female	36	36.0
Educational Level		
Primary	**54**	**54.0**
Preparatory	46	46.0
Child's rank		
First	60	**60.0**
Second	23	23.0
Third	14	14.0
Fourth	3	3.0
Residence		
Urban	59	**59.0**
Rural	41	41.0

Table (2): Frequency distribution of the studied children according to their medical history (n=100).

Medical History	No.	%
Age at onset of disease		
Since birth	12	12.0
In infanthood	83	**83.0**
School age	5	5.0
Adolescent age	0	0.0
***Tests were done to diagnose child condition**		
Complete blood count	96	96.0
Hemoglobin electrophoresis test	100	**100.0**
***The routine test that the child do**		
Complete blood count	100	**100.0**
Iron percentage	95	95.0
Hemoglobin percentage	100	100.0
Liver and kidney function	92	92.0
Do the child receive blood transfusion?		
Yes	95	**95.0**
No	5	5.0
If yes, times the blood transfused was (n=95)		
Once a week	5	5.2
Once every two weeks	7	7.4
Once every three weeks	72	**75.8**
Once every four weeks	11	11.6
Once every five weeks	0	0.0
Is there are problems while receiving blood cells (n=95)		
Yes	71	**74.7**
No	24	25.3
***If yes, these problems was (n=71)**		
Aching	66	**93.0**
Breathing problem	58	81.7
Rash	61	85.9
Hotness	67	94.4
Have the child ever been enter the hospital		
Yes	80	**80.0**
No	20	20.0
If yes, the reason is (n=80)		
Disease related cause	77	**96.3**
Cause unrelated to the disease	3	3.8
***If the reason is related to the disease, what is it? (n=77)**		
To receive treatment of the disease	74	**96.0**
To treat complication of the disease	48	62.3
Is the child committed with iron chelation therapy?		
Always	94	**94.0**
Sometime	6	6.0
Never	0	0.0

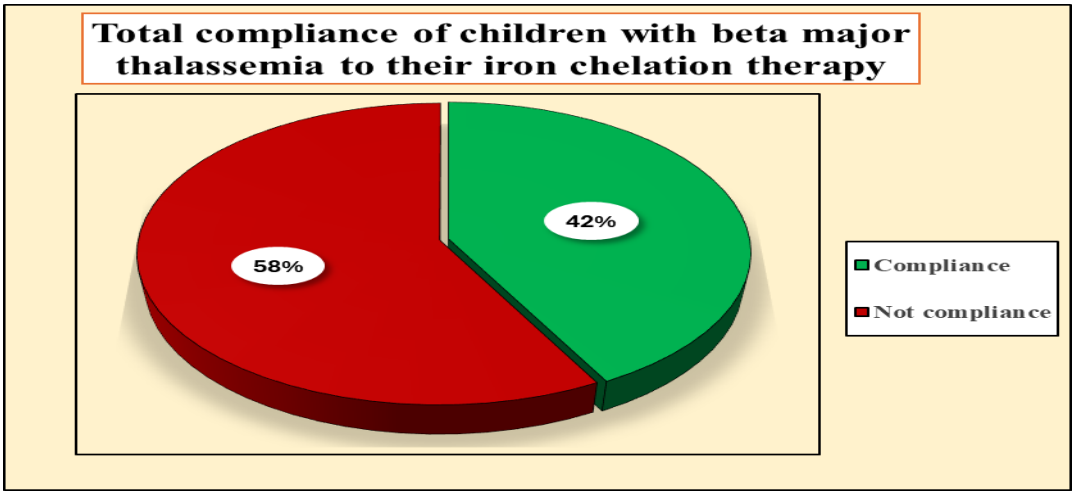

Figure (1): Distribution of the studied children according to their total compliance to iron chelation therapy (n=100).

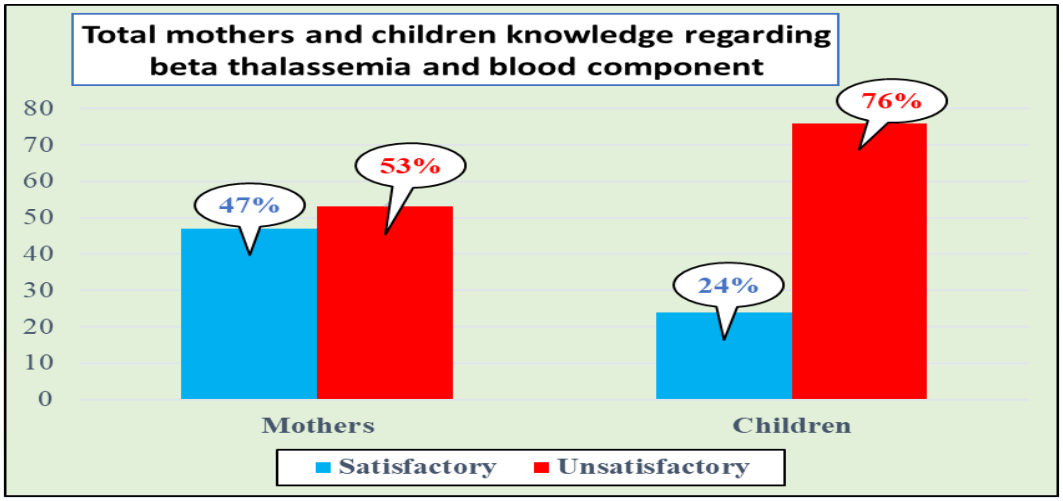

Figure (2): Distribution of the studied mother and children according to their total level of knowledge regarding blood component and beta thalassemia (n=100).

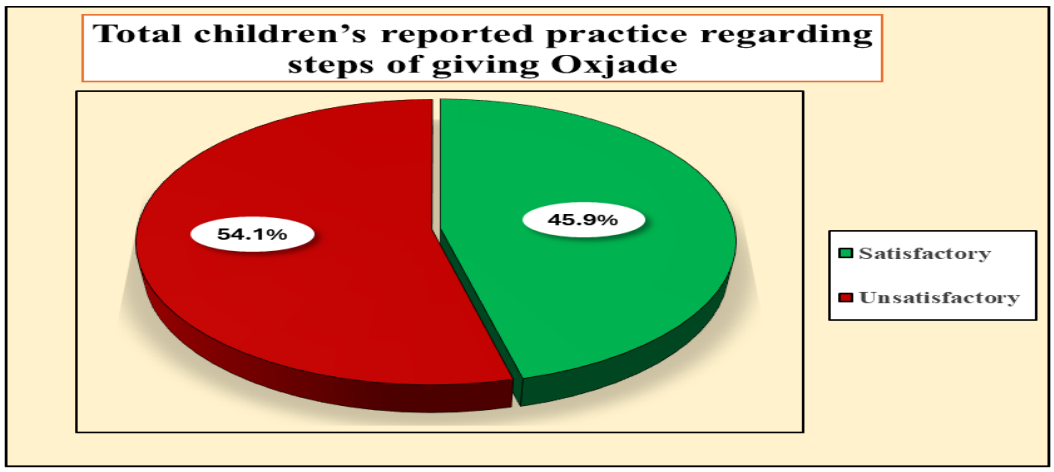

Figure (3): Distribution of the studied children according to their total reported practice regarding steps of giving Oxjade (n=74).

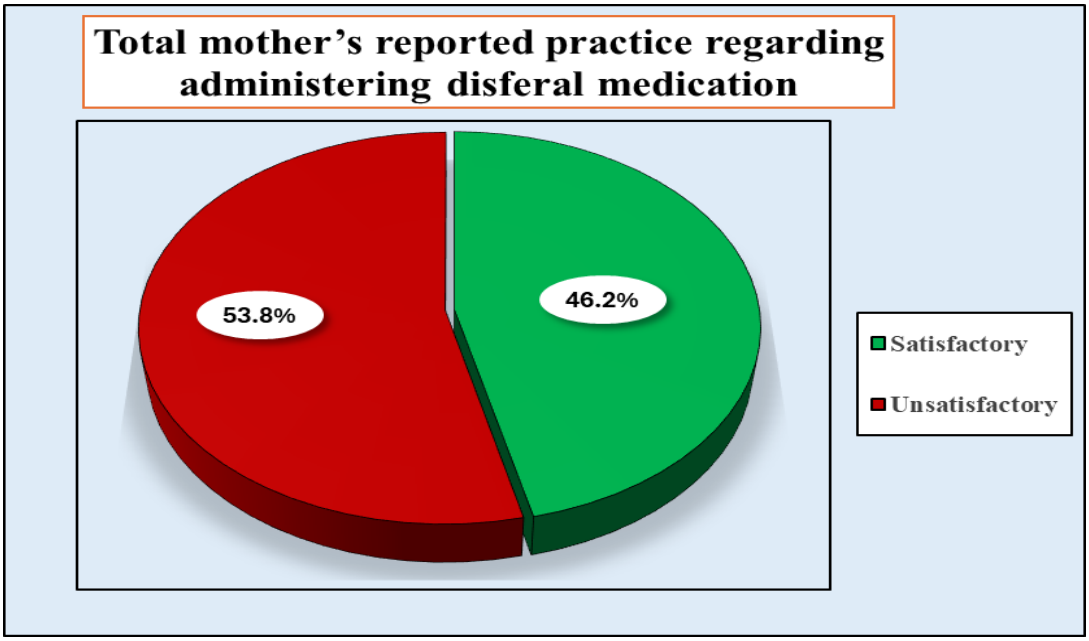

Figure (4): Distribution of the studied mothers according to their total reported practice regarding administering disferal medication (n=26).

Table (3): Relation between personal characteristics of the studied children and total compliance with iron chelation therapy (n=100).

Items		Levels of total compliance with iron chelation therapy				X^2	P-Value
		Compliance (n=42)		Not compliance (n=58)			
		No.	%	No.	%		
Age (years)	6- <9	2	4.8	9	15.5	15.68	0.000**
	9- <12	11	26.2	32	55.2		
	12- <15	29	69.0	17	29.3		
Gender	Male	27	64.3	37	63.8	0.003	0.960
	Female	15	35.7	21	36.2		
Educational Level	Primary	13	31.0	41	70.7	15.48	0.000**
	Preparatory	29	69.0	17	29.3		
Child's rank	First	25	59.5	35	60.4	4.450	0.217
	Second	9	21.4	14	24.1		
	Third	5	11.9	9	15.5		
	Fourth	3	7.2	0	0.0		
Residence	Urban	22	52.4	37	63.8	1.312	0.252
	Rural	20	47.6	21	36.2		

Table (4): Relation between medical history of the studied children and total compliance with iron chelation therapy (n=100).

Medical history of the studied children		Levels of total compliance with iron chelation therapy				X^2	P-Value
		Compliance (n=42)		Not compliance (n=58)			
		No.	%	No.	%		
Age at onset of disease	Since birth	7	16.7	5	8.6	0.843	0.304
	In infanthood	32	76.2	51	88.0		
	School age	3	7.1	2	3.4		
Receive blood transfusion	Yes	39	92.9	56	96.6	0.700	0.647
	No	3	7.1	2	3.4		
History from hospitalization	Yes	34	81.0	46	79.3	0.041	0.839
	No	8	19.0	12	20.7		
Type of iron chelating medication	Desferal	11	26.2	15	25.9	0.001	0.971
	Oxajade	21	73.8	43	74.1		
History from complication after receiving the medication	Yes	8	19.0	8	13.8	0.500	0.479
	No	34	81.0	50	86.2		
Family history from beta major thalassemia	Yes	20	47.6	55	94.8	28.95	0.000**
	No	22	52.4	3	5.2		

Table (5): Correlation between total compliance score, total knowledge score and total reported practices among the studied children (n=100).

Variables	Total compliance score		Total knowledge score	
	r	P-Value	r	P-Value
Total compliance score	**1**	**1**	**1**	**1**
Total knowledge score	0.717	0.000**	**1**	**1**
Total reported practices score	0.903	0.000**	0.610	0.000**

Table (٦): Correlation between total knowledge score and total reported practices among the studied mothers (n=100).

Variables	Total knowledge score	
	r	P-Value
Total reported practices score	0.910	0.000**

Discussion:

Thalassemia is a genetic blood disorder affecting globin chain synthesis with several clinical manifestations relying on the affected globin chain. Beta thalassemia is the most dangerous type that needs regular blood transfusion at an early age. Additionally, those children require iron chelation therapy to overcome iron overload. As a chronic illness, it is considered as great burden not only on the children, but also on the families. This is due to frequent hospital visits for supportive lifelong treatment. Consequently, the principal goal for effective management of thalassemia is to permit children to function with minimal restrictions and enjoy a better life **(Galanello & Origa, 2022).** The present study aimed to Assess the compliance of children with beta major thalassemia to their receiving of iron chelation therapy.

According to characteristics of the studied children, the current study revealed that, less than half of the studied children were in the age group 12-<15 years, with mean age of 12.71±1.71 years. Also, less than two thirds of them were males. Moreover, more than half of them were at primary school. These findings were in an agreement with **(Kannan & Singh, 2021)** who conducted a study about "Compliance score as a monitoring tool to promote treatment adherence in children with thalassemia major for improved physical growth" and reported that one third of the study sample aged between 1-15 years with mean age (10.18 ± 4.98), and less than two thirds of them were males.

On the other hand, these findings were in contrast with **(Keowmani et al., 20٢٣)** who conducted a study about "Adherence to Iron Chelation Therapy Among Children with Beta Thalassemia Major: A Multicenter Cross-Sectional Study" who reported that about half of children were females aged between 6 and 12 years old, and less than half of them attending primary schools.

As regards birth order and residence of the studied children, the result of the present study illustrated that less than two thirds of them were the first child between their siblings. In addition, more than half of them reside in urban areas. This finding disagreed with **(Mostafa & Ab Elaziz, 2014)** who conducted a study about "Factors affecting compliance plan of thalassemic children and their mothers in Outpatient Clinic" and reported that more than half of the studied children were second and third birth order and living in rural areas.

According to medical history the studied children, the result of the present study presented that, the majority of the studied children had a disease from infanthood. Also, all of them had a hemoglobin electrophoresis test to diagnose the disease and had a complete blood count as a routine. This finding was in accordance with **(Keowmani et al., 2023)** who reported that the age at diagnosis of thalassemia in most of the sample was at one year old (infanthood).

According to blood transfusion, the vast majority of the studied children receive blood transfusion, three quarters of them receive once every three weeks. From the researcherss perspectives, it could be due to blood transfusion is the cornerstone for the management of ß-TM. This finding was congruent with **(Mohamed et al., 2022)** who conducted a study entitled "Awareness among parents of children with thalassemia major" and reported that all children had blood transfusions with Inter-transfusion mean interval (days) 10.93±10.96.

Also, the vast majority of them were compliant to iron chelation therapy. Moreover, less than three quarters of them take Oxajade.

Furthermore, the minority of the studied children had a history of complications after receiving the medication, all of them had vomiting and of them had complications that occurred twice after receiving iron chelation therapy. From the researcherss's perspectives, it could be due to chelating therapy to manage and prevent complication associated with ß-TM.

This finding was in the same line with **(Abdel Aziz et al., 20٢٢)** who conducted a study about "Awareness among parents of children with thalassemia major" and reported that all studied children adhered to medication and more than half of them take Oxajade, in addition more than two thirds of them had no complication of treatment. But not similar as regard to type of complication as they revealed that more than half of the studied children had Pain in the joints.

Concerning the studied children's total compliance to iron chelation therapy, the findings of the present study revealed that, more than half of the studied children were not compliance to iron chelation therapy. This study finding disagreed with **(Hossain et al., 2023)** who conduct study about "Health-related quality of life among thalassemia patients in Bangladesh using the SF-36 questionnaire" and found that the percentage of patients who reported good compliance with iron chelating therapy was 95%.

Also, these results were in contrast with **(Mohamed et al., 2022)** who conduct a study entitled -Health-related quality of life among thalassemia patients in Bangladesh using the SF-36 questionnaire- and found that 51.4% had good adherence to iron chelation therapy. The score ranged from 21 to 50, with a mean score of (31.31 ±4.75).

Concerning the studied mother and children total level of knowledge regarding blood component and beta thalassemia, the result of our study revealed that more than half of the studied mother had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. While less than half of them had a satisfactory level of total knowledge. It also shows that, more than three quarters of the studied children had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. While less than one quarter of them had a satisfactory level of total knowledge.

This finding comes in line with that of **(Kharyal et al., 2020)** who carried out a study entitled "Disease Knowledge and General Self-Efficacy among Adolescents with Thalassemia Major and Their Parents' Perspective" and reported that the knowledge of children and their parents about beta thalassemia and blood components is limited and the mean knowledge scores of the adolescent and their parents related to thalassemia were 13.9 ± 2.9 and 13.7 ± 3.6 respectively. Also, these results were in the same line with **Abdel Aziz et al., (2022)** showed that more than half of studied children had unsatisfactory knowledge about ß-thalassemia.

According to the studied children total reported practice regarding steps of giving Oxjade, the current study illustrated that more than half of the studied children had satisfactory practices regarding total reported practice regarding steps of giving Oxjade. While less than half of them had unsatisfactory practices. These findings were supported by **(Haghpanah et al., 2024)** who conduct a study about the "Compliance and satisfaction with deferasirox (oxjade) compared with

deferoxamine in patients with transfusion-dependent beta-thalassemia" and reported that practices were significantly higher among the patients receiving oxjade.

According to the studied mothers total reported practice regarding administering disferal medication, the result of the current study showed that, less than two thirds of the studied children had in satisfactory practices regarding hand washing. While, more than half of them had satisfactory practices regarding administering disferal medication. Moreover, more than half of them had inadequate practices regarding total reported practice regarding steps of administering disferal medication.

This could be explained as a result of the absence of structured educational programs for thalassemic patients and their care givers. These results were consistent with **(Khresheh et al., 2020)** who conduct a study entitled "Knowledge and practices among Mothers about Care of their children with Beta Thalassemia Major: A descriptive Study" and reported that the majority of mothers did not clean their hands correctly and high percentages of mothers practicing the administration of the Desferal incorrectly. Additionally, more than two thirds of mothers had poor scores on the care practice scale.

Regarding relation between personal characteristics of the studied thalassemic children and total compliance with iron chelation therapy, the result of our study presented that, there was highly statistically significant relation between total children's compliance with iron chelation therapy and their age and educational level at P= < 0.01. While, there was no statistically significant relation with their gender, ranking and residence at P= > 0.05.

This finding was supported by **(Trachtenberg et al, 2021)** who conduct study entitled "Iron chelation adherence to deferoxamine and deferasirox in thalassemia" and reported that found that adherence to chelation therapy is better in younger children compared to older children and there was statistically significant relation between age and educational level of the study sample and their total compliance with iron chelation therapy at P= < 0.05.

On the other hand This finding was in contrast with **(Kaur, 2023)** who conduct study about " Adherence to the Iron chelation Therapy among Transfusion Dependent Beta Thalassemic Pediatric Patients-A Cross-Sectional Study" and reported that males tend to have better compliance to chelation therapy also observed a slightly better compliance in the urban families so, there was statistically significant relation between the children's gender and residence and compliance to chelation therapy at P= < 0.05

Concerning correlation between total compliance score, total knowledge score and total reported practices among the studied children, our study indicated that, there were highly significant positive correlation between total children's compliance score and their total knowledge score (r=0.717, p=<0.01), and total reported practices score (r=0.903, p=<0.01). Also, there were highly significant positive correlation between total children's knowledge score and their total reported practices score (r=0.610, p=<0.01) (table, 24).

This finding was agreed with **(Mostafa & Ab Elaziz, 2014)** Who found positive association between treatment adherence and patients' knowledge about their disease.in addition, this finding was supported by **(Kannan& Singh, 2021)** Who found positive correlation between

total children's knowledge score and total reported practices score p=<0.05).

Regarding correlation between total knowledge score and total reported practices among the studied mothers, the result of this study indicated that, there were highly significant positive correlation between total mother's knowledge score and their total reported practices score (r=0.910, p=<0.01). This finding was in contrast with **(Khresheh et al., 2020)** Who found that no significant positive association between the overall level of the knowledge scale about thalassemia of mothers and the overall care practices score of Desferal tablets ($\chi 2$=15.0, (P=0.81).

Conclusion:

More than half of the studied mother had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. While, less than half of them had satisfactory level of total knowledge. Also, more than three quarters of the studied children had unsatisfactory level of total knowledge regarding blood component and beta thalassemia. While, nearly less than one quarter of them have satisfactory level of total knowledge. More than half of the studied children had satisfactory practices regarding total reported practice regarding steps of giving Oxjade. While, less than half of them had unsatisfactory practices. Also, more than half of the studied mothers have in satisfactory practices regarding total reported practice regarding steps of administering disferal medication. While, less than half of them have satisfactory practices. Furthermore, more than half of the studied children are not compliance to iron chelation therapy. While, less than half of them are compliance to iron chelation therapy.

Recommendations:

- Emphasize the role of nurse in education the thalassemic children and their mothers regarding compliance to iron chelation therapy.
- Emphasize the role of nurse in Encouraging thalassemic children and their mothers to participation in recreation activities to enhance their quality of life.
- Availability of designed guidelines in Arabic language regarding care of children with beta major thalassemia in different health care settings.
- Provide practical knowledge to children with thalassemia major and their parents is mandatory to significantly improve children compliance to treatment.
- The pediatric health nurses should be specially trained to act as genetic counselors to reduce the incidence of thalassemia and should be able to discuss important aspects of prevention.

Recommendations for further researches: Conducting the study in different settings of pediatric hematologic clinics to generalize the results of the study and raise the level of awareness about the importance of compliance of beta major thalassemia children to iron chelation therapy.

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التزام الأطفال المصابين ببيتا ثلاسيميا الكبرى بتلقيهم علاج سحب الحديد

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بيتا ثلاسيميا الكبرى هي اضطراب وراثي في الهيموجلوبين ينتج عن غياب سلسلة بيتا غلوبين في مسار إنتاج الهيموجلوبين. ويتطلب نقل الدم بانتظام مما يؤدي إلى زيادة الحديد مما يتطلب العلاج بسحب الحديد لذلك هدفت الدراسة إلى تقييم التزام الأطفال المصابين ببيتا ثلاسيميا الكبرى بتلقيهم علاج سحب الحديد، وقد تم استخدام تصميم بحث وصفي لإجراء هذه الدراسة، و أجريت الدراسة في مبنى الباطنة بالدور الثاني في وحدة أمراض الدم في مستشفى بنها التخصصي للأطفال التابع لأمانة المركز الطبي المتخصص، و قد شملت الدراسة على عينة غرضية مكونه من ١٠٠ طفل تم تشخيصهم ببيتا ثلاسيميا الكبرى وأمهاتهم اللائي حضرن المكان المذكور سابقًا، و قد أسفرت النتائج على أن الغالبية العظمى من الأطفال الذين أجريت عليهم الدراسة لا يلتزمون بعلاج سحب الحديد. كما أن غالبية الأمهات اللاتي أجريت عليهن الدراسة لديهن مستوى غير مرضي من المعرفة فيما يتعلق بمكونات الدم وبيتا ثلاسيميا. كما أن أكثر من ثلاثة أرباع الأطفال الذين أجريت عليهم الدراسة لديهم مستوى غير مرضي من المعرفة فيما يتعلق بمكونات الدم وبيتا ثلاسيميا. وعلاوة على ذلك، فإن أكثر من نصف الأطفال الذين أجريت عليهم الدراسة لديهم ممارسات مرضية فيما يتعلق بإجمالي الممارسة المبلغ عنها فيما يتعلق بخطوات إعطاء الأوكسجيد. وأكثر من نصف الأمهات اللاتي أجريت عليهن الدراسة لديهن ممارسات مرضية فيما يتعلق بإجمالي الممارسة المبلغ عنها فيما يتعلق بخطوات إعطاء دواء الديسفرال، وقد لخصت النتائج أن أكثر من نصف الأطفال الذين أجريت عليهم الدراسة لا يلتزمون بعلاج سحب الحديد. في حين أن أقل من نصفهم يلتزمون بعلاج سحب الحديد، و قد أوصت الدراسة على ضرورة التأكيد على دور الممرضة في تثقيف الأطفال المصابين بالثلاسيميا وأمهاتهم فيما يتعلق بالالتزام بعلاج سحب الحديد.