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 led to iron overload that require iron chelation therapy. **Aim of study:** was to assess the compliance of children with beta major thalassemia to their receiving of iron chelation therapy. **Design:** A descriptive research design was utilized to conduct this study. **Setting:** The study was conducted at outpatient clinic in hematological diseases Unit at Banha Specialized Pediatric Hospital affiliated to secretariat of specialized medical center. **Subject:** 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 compiling to iron chelation therapy. While, less than half of them are compiling to iron chelation therapy. **Recommendations:** Emphasize the role of nurse in educating the thalassemic children and their mothers regarding compliance to iron chelation therapy.

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

Among the many frequent monogenic hereditary haemoglobin diseases is beta major thalassaemia, often known as Cooley anaemia. The production of haemoglobin is interrupted because a beta-globin chain is missing from the process. A genetic condition known as beta-thalassemia causes either a reduction in beta-globin chain size or its complete absence due to one of over 200 different point mutations in the beta-globin gene. The main cause of the disease is inefficient red blood cell production. Consequently, children affected by thalassaemia develop a lifelong reliance on blood transfusions. The high rates of illness and death caused by this disorder may be prevented with early detection and treatment (Khan

& Shaikh, 2023).

The most common monogenic illness in the world is thalassaemia, a genetic ailment that causes irregularities in the quantity of haemoglobin. Carriers of thalassaemia genetic mutations are believed to constitute 1.5-7% of the global population. Southeast Asia, the Indian subcontinent, the Middle East, portions of North and sub-Saharan Africa, and the Mediterranean are the areas where thalassaemia is most common. A worldwide health problem is the appearance of these illnesses in big multiethnic cities in North America and Europe as a result of continuous migration (Li, et al., 2024).

Based on the purpose of chelation, deferoxamine was shown to be an effective intramuscular chelation agent; however, it was found to be much more effective when delivered as a subcutaneous infusion over 8 to 24 hours. Side effects of subcutaneous infusions include soreness and oedema at the injection site, which may occur when the needle is injected into subcutaneous tissue on the legs or belly. Deferirox does not It may be taken orally once day and can be used alone or in conjunction with the other two medications. Symptoms such as stomach discomfort, vomiting, and diarrhoea are common adverse effects. You may get Deferiprone in pill and syrup forms, and the recommended dosage is three times daily. Arthralgias, a low white blood cell count, and gastrointestinal side effects such nausea, vomiting, and diarrhoea are common side effects (Eziefula, et al., 2022)

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Aim of the study

Examining how well iron chelation treatment is adhered to by children with beta major thalassaemia was the primary goal of this research.

What percentage of youngsters undergoing iron chelation treatment for beta major thalassaemia really take all of their prescribed medication as prescribed?

Subjects and Methods

Subjects and Methods: This research set out to determine how well iron chelation treatment was adhered to by children suffering with beta major thalassaemia.

The present study's subjects and methodologies will be covered under three different designs:

Technical blueprinting.

Design for operations.

Management planning. IV. Methods of statistical analysis.

Architecture from a technical perspective:

The technical design included the following elements: study design, context, participants, and data gathering instruments.

Methodology of the inquiry:

This research made use of a descriptive quantitative study design.

Setting: The research was place at Banha Specialised Paediatric Hospital, which is linked with the secretariat of a specialised medical centre, in an outpatient clinic inside the haematological disorders unit.Fields of study:

Inclusion criteria for the 100 children with beta major thalassaemia and their mothers who were part of the purposive sample and who were present in the aforementioned setting were:

Kids' ages vary from six to fifteen.

Absolutely no additional long-term health issues. 3-Getting treatment with iron chelation.

Data Collection Instruments:

This tool was used to gather data:

First Aid: A Questionnaire for Organised Interviews (Appendix)

Elsoudy et al. (2022) and Mahmoud (2019) both used this instrument, which the researcher had created and amended with the help of supervisors after consulting

relevant literature. Iron chelation treatment compliance was evaluated in children with beta major thalassaemia using this tool. For the sake of the mother and children's comprehension, it was written in a straightforward form of Arabic. The following were its five components:

Individual Features of Children Diagnosed with Beta Major Thalassaemia (Part 1)

In this section, we looked at the individual traits of kids who have beta major thalassaemia. There are five questions covering the child's age, gender, educational background, rank, and place of residence.

Part 2: - Mothers' Individual Traits in Relation to Their Children's Beta Major Thalassaemia

The purpose of this section was to evaluate the adherence of beta major thalassaemia patients to their iron chelation treatment. Elsoudy et al. (2022) provided the basis for its adoption. The five primary components were as follows: Whether or not a child is dedicated to eating healthily, which includes cutting back on iron-rich foods like aubergine and liver, increasing consumption of iron-absorbing foods like oranges, lemons and green peppers, cutting back on foods with a high percentage of preservatives, like chips and Pepsi, and eating plenty of folic acid-containing foods, like okra and lettuce. The second item is the level of pharmaceutical commitment on the part of the kid. This includes four sub-items, such as taking the medicine exactly as given, getting immunisations on a regular basis to avoid infection, and taking antibiotics as directed. The third item is the compliance of children with thalassaemia with blood transfusions. This entails four sub-items: going to the hospital for transfusions on a regular basis, doing what the doctor says to do everyday after a transfusion, eating what the doctor says to eat, and telling the doctor right away if anything goes wrong. The fourth item is for children with thalassaemia to comply with personal hygiene practices. This includes seven sub-items, such as: frequently washing hands, clipping nails, taking care of hair, bathing daily, cleaning teeth regularly, and using the child's own tools and not sharing them with a sibling. Fifth item: Encouraging Children with Thalassaemia to Follow Up: this item has six subitems, including: going to the doctor for checkups on a regular basis, eating according to the doctor's orders, keeping track of weight, getting regular dental checkups, telling doctors about the child's condition before any tests or surgeries, and going to the doctor right away if their health starts to worsen.

The method for evaluating the compliance of children:

On a three-point Likert scale, each item would be assessed as follows: The following scores were assigned: (2) for always complying, (1) for occasionally complying, and (0) for never complying.

We computed and categorised the entire score system for the child's compliance as follows:

Complete conformity was achieved (75–100%)

A score of 60% to 75% was achieved with some compliance.

- The compliance score was never between 60% and 50%.

RESULTS

Part I: **Individual traits of the moms and children who were part in the research.**

Statistical distribution of the 100 children included in the study, broken down by demographic variables (Table 1).

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 (1): The results demonstrate that the children surveyed fell within the age bracket of 12-<15 years, with an average age of 12.71±1.71 years, constituting less than half of the total. Regarding the gender breakdown, men make up fewer than two thirds (64.0%) of the total. Additionally, 54.0% of them were elementary school students. Plus, little under 60% of them were the firstborn out of many siblings. Not to mention that 59.0% of them call cities home.

Table (2): Statistical distribution of the moms whose features were investigated (n=100).

Personal characteristics	No.	%
Age (years)		
25 - <30	5	5.0
30 - <35	22	22.0
35 - <40	56	**56.0**
≥ 40	17	17.0
Mean ± SD	**36.96±3.41**	
Educational level		
Read and write	6	6.0
Primary	7	7.0
Preparatory	10	10.0
Secondary	51	**51.0**
University	26	26.0
Job		
Working	16	16.0
House wife	84	**84.0**
Residence		
Urban	59	**59.0**
Rural	41	41.0
Marital status		
Married	93	**93.0**
Divorced	4	4.0
Widow	3	3.0
Family income		
Enough	41	41.0
Not enough	59	**59.0**
Is there consanguinity relation between the father and the mother?		
Yes	65	**65.0**
No	35	35.0
***If yes, what is the degree of Consanguinity? (n=65)**		
Blood Consanguinity	44	**67.7**
Pedigree Consanguinity	8	12.3
First degree relative	5	7.7
Second degree relative	8	12.3
Family history from beta major thalassemia		
Yes	75	**75.0**
No	25	25.0
***If yes, who is the person suffer from the disease? (n=75)**		
Father	60	**80.0**
Mother	53	70.7
Brother	8	10.7
Uncles	4	5.3
Grand parents	2	2.7
Disease condition		
Carrier of the disease	71	**94.7**
Has the disease	4	5.3

(*) Responses not mutually exclusive. **SD=** Standard deviation.

Table (2): It is shown that the majority of the moms surveyed (56.0%), with an average age of 36.963.41 years, fall within the age bracket of 35-40 years. Over half of them (51.0%) also held a secondary degree. Not only that, but 84 percent of them are ineffective. Not to mention that 59.0% of them call cities home. Additionally, 93.0% of them are in committed marriages. Additionally, 59.0% of them are underemployed. In addition, whereas almost two-thirds of the mothers surveyed had blood consanguinity, nearly two-thirds (65.0%) had consanguinity relations with their husbands. In addition, 75.0% of the moms surveyed had a history of beta major thalassaemia in their family, 80.0% of those mothers are fathers, and 94.7% of those mothers are carriers.

Part (II): Medical history of the examined youngsters.

Table (3): Medical history-based frequency distribution of the 100 children included in the study.

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, how many times the blood transfused? (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, what these problems? (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, what is the reason? (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

Table (3): carried on...

Medical History	No.	%
Type of iron chelating medication that the child takes		
Desferal	26	26.0
Oxajade	74	**74.0**
Is there are any complications after receiving the medication?		
Yes	16	**16.0**
No	84	84.0
If yes, what this complication? (n=16)		
Vomiting	16	**100.0**
Diarrhea	7	43.8
Dizziness	13	81.3
Number of times the complication occurred after receiving iron chelation therapy (n=16)		
Once	3	18.7
Twice	13	81.3

(*) The answers are not mutually complimentary.

Part (III): **Evaluation of iron chelation treatment adherence in children diagnosed with beta major thalassaemia.**

Distribution of the examined children's frequencies according to their adherence to the right nourishment and prescribed medicine (n=100) is shown in Table (4).

Items	Always		Sometimes		Never	
	No.	%	No.	%	No.	%
Compliance to the proper nutrition						
Reducing iron rich foods such as eggplant and liver	15	15.0	67	67.0	18	18.0
Eat food that help absorption of iron, such as orange, lemon and green pepper	21	21.0	70	**70.0**	9	9.0
Reducing foods that contain high percentage of preservative, such as chips and Pepsi	29	29.0	56	56.0	15	15.0
Eat large amounts of foods that contain folic acid, including lettuce and okra	20	20.0	68	**68.0**	12	12.0
Compliance to the prescribed medication						
Take the right prescribed dose	51	**51.0**	46	46.0	3	3.0
Take vaccinations regularly to prevent any infection	53	**53.0**	44	44.0	3	3.0
Receiving preventive antibiotics according to doctor order.	50	50.0	42	42.0	8	8.0

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ISCUSSION

Multiple clinical presentations are dependent on the afflicted globin chain in thalassaemia, a hereditary blood condition that affects globin chain production. The most deadly form of the blood disorder, beta thalassaemia, requires frequent transfusions beginning at a young age. Iron chelation treatment is also necessary for such kids to get rid of their iron excess. Both the children and their family bear the brunt of this chronic condition. This is because patients need ongoing supportive therapy, which requires repeated hospital visits. Therefore, allowing children to live healthier lives with fewer constraints is the primary objective of successful thalassaemia care (Galanello & Origa, 2022).

Examining how well iron chelation treatment is adhered to by children with beta major thalassaemia was the primary goal of this research.

The present research found that, based on the kids' traits, just about half of the kids surveyed fell into the 12-~15 age bracket, with an average age of 12.71±1.71 years. Plus, there were less than 70% men among them. Table 1 also shows that almost half of them were in elementary school. The results were in line with those of a study by Kannan and Singh (2021) titled "Compliance score as a monitoring tool to promote treatment adherence in children with thalassaemia major for improved physical growth." The researchers found that a third of the participants were between the ages of 1 and 15, with a mean age of 10.18 4.98, and that fewer than two thirds of the participants were male.

Contrarily, in a study titled "Adherence to Iron Chelation Therapy Among Children with Beta Thalassaemia Major: A Multicenter Cross-Sectional Study" (Keowmani et al., 2023), it was found that less than half of the children, who were between the ages of 6 and 12, were attending primary school, and that approximately half of the children were female.In terms of birth order and residency, the current research found that less than two thirds of the children were the firstborn out of multiple siblings. Furthermore, almost 50% call cities home (table, 1). More than half of the children analysed in a 2014 research by Mostafa and Ab Elaziz on "Factors affecting compliance plan of thalassemic children and their mothers

in Outpatient Clinic" were of second or third birth order and lived in rural regions; this conclusion contradicts that finding

Conclusion

We inferred from the results of the present investigation that:

In terms of overall understanding on blood components and beta thalassaemia, almost half of the mothers surveyed had an inadequate level. As a result, just over 50% of them possessed enough general knowledge. Additionally, the overall level of knowledge of blood components and beta thalassaemia was unacceptable among almost three quarters of the youngsters surveyed. However, when it comes to overall knowledge, about 25% of them fall short. Additionally, in terms of overall reported practice about stages of providing Oxjade, over half of the children evaluated had good practices. Despite this, little under 50% of them engaged in underwhelming procedures. In addition, when it comes to the entire stated practice regarding the processes of delivering ferral medicine, over half of the moms evaluated had good practices. But only about 50% of those people really have good habits. Iron chelation treatment is also not adhered to by over 50% of the youngsters surveyed. Iron chelation treatment is followed by less than 50% of patients.

Recommendations

Based on the results of this investigation, we propose the following:

Highlight the importance of the nurse's involvement in educating mothers and children with thalassemic conditions on the need of adhering to iron chelation treatment.In order to improve their quality of life, encourage moms and children with thalassemic to participate in recreational activities.

Healthcare facilities that provide treatment to children with beta major thalassaemia have access to Arabic-language guidelines for this condition.

In order to greatly enhance treatment compliance among children with thalassaemia major, it is essential to provide them and their parents with practical information.

In order to lower the thalassaemia incidence, paediatric health nurses should have specialised training to advise patients on genetic testing and

other preventative measures.

Further research should be carried out in other paediatric haematologic clinics to generalise the study's findings and increase awareness of the significance of iron chelation treatment compliance in children with beta major