Knowledge of Children and Adolescents with Beta Thalassemia Major about their Disease: An Assessment Study

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Abstract

Background: Beta-thalassemia major is the most common hemoglobin disorder in the world. Assessing the knowledge of the children and adolescents regarding a beta-thalassemia major is important because helps to avoid complications. This study **aimed** to assess knowledge of children ß-TM about their disease. **Design:** Descriptive exploratory design. **Settings:** The study conducted at Hematology Outpatient Clinics and inpatients word at Children's Hospital affiliated to Ain Shams University. **Sampling:** A purposive sample comprised of 80 children with beta thalassemia major. **Tools: Part 1:** Designed questionnaire. **Part 2:** History of studied children regarding beta-thalassemia major. **Part 3:** Knowledge of the children regarding beta-thalassemia major. **Part 4**: Evaluating informational needs of studied children about beta-thalassemia major. **Results:** Current study revealed that the mean age of studied children was 14.25 ± 2.8 years and slightly more than half of them were female, about more than half of studied children had unsatisfactory knowledge about beta-thalassemia major. **Conclusion:** Current study revealed that more than half of the studied children were had unsatisfactory knowledge regarding beta-thalassemia major. **Recommendation:** Provide continuous health education to children with beta-thalassemia major regarding their disease.

Keywords: Beta thalassemia major – Children – Adolescents– Knowledge.

Introduction:

Beta-thalassemia major (ß-TM) is the most common hemoglobin disorder in the world and thalassemia major stand among the most severe forms ***(Abu-Muammar, 2020).*** Despite the life-saving nature of long-term blood transfusion, iron toxication due to dysregulated cellular iron metabolism is the leading cause of prolonged complications in children and adolescents with Beta-thalassemia major ***(Chuansumrit et al., 2017).***

Knowledge about the disease can prevent disease related complication and improve quality of life. Beta-thalassemia major can easily be prevented by knowledge, education, screening, premarital genetic counseling and prenatal diagnosis ***(Vasudeva Murthy et al., 2015; Haque et al., 2015).*** Many factors affect the knowledge of transmission of disease and option of preventive services ***(Hanprasertpong et al., 2018).***

Nurses play a critical role in managing children with Beta-thalassemia major by helping to prevent unnecessary complications and providing treatment aimed at minimizing interference with the demands of the daily life. This includes e.g. instructing the children and their family about the detection and reporting the serious symptoms such as fever or pain, clarifying the importance of cleanliness and encouraging interaction with other health professionals ***(Azize, Tahir & Kelsey, 2015).***

Significance of the Study

Until now children with ß-TM and their families face many challenges. Health related lifestyle aspects and related physical and psychological problems of children and adolescents with ß-TM were dim and ignored.

Beta thalassemia major is a major public health problem in Egypt with particularly high incidence due to strong cultural preference for consanguineous marriages ***(Adly & Ebeid, 2015).*** It has been estimated that 1000 children out of 1.5 million live births are born annually with thalassemia major; in multicenter studies, the carrier rate in Egypt was reported to range from 5.3 to ≥9% and a gene frequency of 0.03 ***(Sherief et al., 2014; El-Shanshory et al., 2014; Elmezayen et al., 2015).***

The aim of the study:

This study aims to assess knowledge of children with ß-TM about their disease.

Research Design:

A descriptive design was utilized to conduct the study.

Technical design

The technical design was included research setting, subjects and tools for data collection.

Research Setting:

This study conducted at Hematology Outpatient Clinics at Children's Hospital affiliated to Ain Shams University which found in the 4th floor. There is a caravan for blood transfusion in the ground floor. In addition to 3 inpatients wards in the 2nd floor in the mentioned hospital.

Research Subjects:

A purposive sample of children and adolescents was selected according to certain inclusion criteria. The sample size was determined statistically by power analysis considering the total number of ß-TM children and adolescents in the previously mentioned setting along 2 years from 2015 till 2017. Accordingly, the sample size was 80 children.

Inclusion Criteria:

The study subject was selected according to the following inclusion criteria:

1. Children and adolescents with ß-TM aged 9 years to 18 years.
2. Children and adolescents have long duration of ß-TM (9 years & more).

Tools for data collection:

Data was collected using the following Tools:

Designed questionnaire:

It was developed in a simple Arabic language by the researcher after reviewing the related literatures and reviewed by supervisors; it was consisted of four parts:

Part 1: It concerned with characteristics of the study subjects, as follows:

a) Characteristics of the studied children and adolescents including: Age, gender, education level, ranking and residence.
b) Characteristics of caregivers including: Relation, age, level of education and working.

Part 2: Medical history of studied children about ß-TM.

Part 3: It concerned with knowledge of the children about; ß-TM it composed of (26) questions to assess the level of children with ß-TM knowledge regarding ß-TM.

Part 4: It concerned with evaluating informational needs of studied children about ß-TM which composed of (12) questions.

Operational design:

Field Work

The actual field work was carried out over a period of 5 months from beginning of January 2019 up to the end of May 2019. The researcher was available in the study setting 2 days (Saturday and Sunday)/week by rotation according to children with ß-TM

appropriate time to collect data. From 9 am to 1 pm. The children were interviewed (for 45-60 minutes). The researcher started the interview by introducing herself to the children, giving them clear and brief idea about the aim of the study and its expectation then starting the interview questionnaire and each child with ß-TM was interviewed individuals to answer the questions.

Validity:

Tools of the present study were judged by a panel of 3 expertise and they were professors of pediatric nursing. The necessary modifications were done according to experts' opinion to ensure validity of the content.

Reliability:

The reliability for tool was 0.74.

Using Alpha Cronbach Reliability Analysis of the Used Tool

Exploratory phase:

A pilot study was carried out during November 2018 involved 10% (8 of children with beta thalassemia major) of the total study sample. The result of the data obtained from the pilot study helped in removing of some repeated questions related to knowledge to avoid duplication of questions and then all children involved in the pilot study were included of the study sample.

Administrative design:

Approval was obtained through an issued letter from Dean of Faculty of Nursing, Ain Shams University to the Hospital Director of the previously mentioned setting and the researcher was explaining aim of the study and its expected outcomes.

Statistical Design:

The data obtained was organized, analyzed, and presented in the form of tables and figures using the Statistical Package for Social Sciences (SPSS) Version 20. Qualitative variables were presented in the form of frequencies and percentages; quantitative variables were presented in the form of mean and SD. Qui square and Fishers Exact tests were used to test the significance of results obtained. Statistical significant difference was considered at $P < 0.05$.

Ethical considerations:

The ethical considerations in the study included the following:

All the gathered data was used for research purpose only. The study sample was informed about the purpose and expected outcomes of the study and they was assured that the study is harmless and their participation is voluntary and they have the right to withdraw from the study at any time and without given any reason. They were assured also that anonymity and confidentiality were guaranteed.

Results:

Table (1): shows that, more than two fifth (42.5%) of the studied children aged from 13 to <16 years with a mean age 14.25±2.84 years and more than half (57.5%) of them were females. Moreover, this table reveals that, two fifths (40%) of the studied children had primary education, less than two thirds (62.5%) of them were from rural areas.

Table (2): illustrates that all (100%) of studied children taken medication and blood transfusion as treatment and more than three quarters (78.6%) of them know sites of desferal injection.

As observed from this table more than half (57.5%) of studied children had correct knowledge about medical treatment of ß-TM Moreover, the side effect of treatment was unknown for half (50%) of the studied children.

Table (4): reveals that half (50 %) of studied children had correct knowledge about the best action in occurring side effect

from blood transfusion and two fifth (40%) of them had correct knowledge about foods that increasing the iron in blood.

Figure (1): Shows that more than half (56.2%) of studied children had unsatisfactory knowledge about ß-TM.

Figure (2): reveals that more than half (51.25%) of studied children and adolescents with ß-TM had information needs regards ß-TM.

Table (1): Distribution of the studied children with ß-TM according to their characteristics (n=80).

Characteristics of children		No.	%
Age (years)			
9 < 12		16	20
12 < 15		34	42.5
15≤18		30	37.5
Mean±SD	14.25±2.84		
Gender			
Male		34	42.5
Female		46	57.5
Level of education			
Primary		32	40
Preparatory		26	32.5
Secondary		22	27.5
Ranking			
First		23	28.7
Second		16	20
Third		16	20
Fourth		25	31.3
Residence			
Urban		30	37.5
Rural		50	62.5

Table (2): Distribution of the studied children with ß-TM according to their present history related to medications and complications (n=80).

present history of medications	No.	%
Types of treatment		
Medication	80	100
Blood transfusion	80	100
Types of medication		
Desferal	28*	35
Ferriprox	11	21.2
Exjade	41	78.8
mixed mediation	72	90
Method of taking desferal injection	N= 28	
Intravenous	10	35.7
Subcutaneous infusion	18	64.3
Sites of desferal injection		
Know	22	78.6
Not know	6	21.4
Times of desferal taken		
Five times a week	12	42.9
Once a week	6	21.4
Once a month	10	35.7
Side effects of desferal		
Puffiness and redness in the injection area	18	64.3
Black or reddish urine or stool	2	7.2
Vision disorder and blurred Vision	8	28.5
Dealing with side effects of desferal		
Visit physician	22	78.6
Stop treatment if side effect occur	6	21.4
Complications of treatment		
Yes	80	100
If yes, list it		
Pain in the joints	17	21.2
Problems in the vision	9	11.3
No complication	54	67.5

*Number are not mutual exclusive

Table (3): Distribution of the studied children with ß-TM according to their knowledge about beta thalassemia major (n=80).

Knowledge about ß-TM	Correct		Incorrect	
	No.	%	No.	%
Definition	38	47.5	42	52.5
Risk factors	28	35	52	65
Types	32	40	48	60
Signs and symptoms	52	65	28	35
Diagnosis	56	70	24	30
Normal range of iron in blood	32	40	48	60
Level of hemoglobin in children	18	22.5	62	77.5
Level of hemoglobin in adolescence	25	31.3	55	68.75
Complications	40	50	40	50
Medical treatment	46	57.5	34	42.5
Side effects	40	50	40	50
Prevention	30	37.5	50	62.5
Self care	34	42.5	46	57.5

Table (4): Distribution of the studied children with ß-TM according to their knowledge about blood transfusion and iron condition (n=80).

Knowledge about blood transfusion and iron condition	Correct		Incorrect	
	No.	%	No.	%
Importance	32	40	48	60
Hazards if blood transfusion not given	38	47.5	42	52.5
Side effects occur during or after blood transfusion within 24 hours	46	57.5	34	42.5
The best action when side effect from blood transfusion	40	50	40	50
Complications of recurrent blood transfusions	56	70	24	30
Causes of complications	30	37.5	50	62.5
Foods that increasing the iron in blood	32	40	48	60
Foods or drinks decrease iron absorption	44	55	36	45
Damages of increase percentage of iron in the blood	36	45	44	55

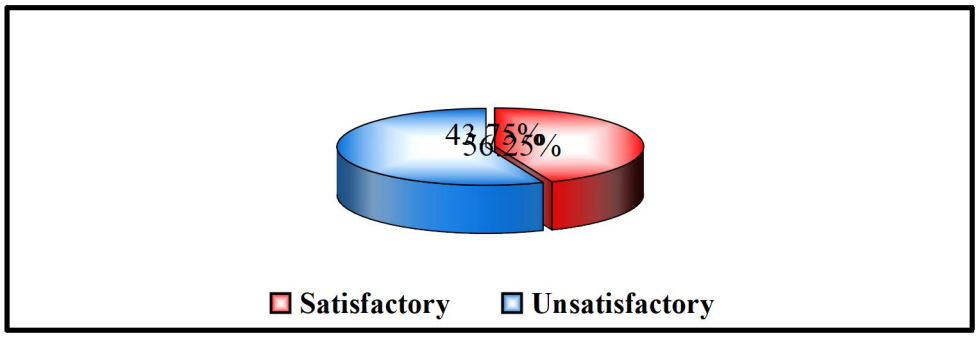

Figure (1): Percentage distribution of the studied children with ß-TM according to their total knowledge about beta thalassemia major (n=80).

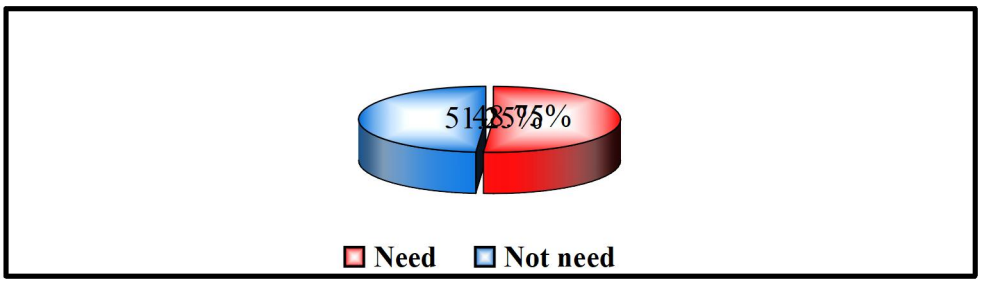

Figure (2): Percentage distribution of the studied children with ß-TM according to their total informational needs (n=80).

Discussion:

Beta thalassemia major is the commonest inherited hemolytic disease in the world, due to disruption of beta globin biosynthesis. Children with ß-TM present along a spectrum ranging from nearly asymptomatic to severe anemia requiring lifelong blood transfusions ***(Benz & Angeluci, 2018).***

Lack of knowledge and education about ß-TM is a major non-biomedical factor, which leads children to face serious social, economic, and psychological problems ***(Ebrahim et al., 2019).***

Pediatric nursing for the child with ß-TM is centered on family teaching & family support as well as actual treatment of the anemia. The hematology nurse is

responsible for administering blood transfusions and observing for transfusion reactions ***(Zorac et al., 2013).***

Regarding to characteristics of the study subjects, the findings of the present study showed that the mean of the studied subject was 14.25±2.84 years. This finding was contrary to that of ***Gharaibeh et al., (2018)*** who mentioned in a study entitled "Clinical Burdens of ß-TM in Affected Children" that more than half of children ages less than 12 years and they revealed that more than half were females. This finding was in agreement with that of ***Khan et al., (2019)*** who carried out a study entitled "Quality of Life of Thalassemia Major Patients" and found that the majority of the studied subject was females. Meanwhile this finding was contrary to the finding of ***Hakeem et al., (2018)*** who carried out a study entitled "Health-Related Quality of Life in Pediatric and Adolescent Patients with Transfusion Dependent ß-Thalassemia in Upper Egypt (Single Center Study)" and found that most of the study subject was males.

The findings of the present study showed that two fifth of the studied children were preparatory school. This finding disagreement with ***Hisam et al., (2018)*** who carried out a study entitled "Perceived Stress and Monetary Burden among Thalassemia Patients and their Caregivers" and reported that regarding education status of studied subject having more than two fifth primary educations. Moreover, this finding was not supported by ***Ali, El-Bilsha & Mohamed, (2018)*** who carried out a study entitled "Coping Strategies among Children with Thalassemia" and mentioned that more than half of the studied thalassemic children not entered school.

The results of the present study revealed that slightly less than two thirds of the studied children from rural area. This could be attributed to the fact that rural areas face a problem of deficient quality health care services and health care providers. This shed the light on the importance of strengthening the rural areas with proper health care services in order to fulfill their residences needs of effective and efficient health services. The same results could be attributed to the high prevalence of poverty and illiteracy among rural population. This finding was in accordance with ***Gharaibeh et al., (2018)*** who mentioned that less than two thirds of children from rural area. However, these findings were similar to a study done by ***Abusaad & Sarhan, (2016)*** who carried out a study entitled "Exercise Training Program and Tele nursing Effects on Depression and Fatigue Level in B-Thalathemia Major Children", this study revealed that most of studied children were from rural areas.

Additionally, the findings of the current study revealed that, all of studied children had medication and blood transfusion treatment. It could be due to the blood transfusion is the cornerstone for the management of ß-TM and chelating therapy to manage and prevent complication associated with ß-TM. This findings were agreed with ***Bhojak et al., (2020)*** who carried out a study entitled "Efficacy of Once a Month Single Dose Intravenous (Deferoxamine) versus Daily Oral (Deferasirox) Iron Chelator in Thalassemia Major: An Open Label Randomized Parallel Group Active Control Interventional Trial" and found that all patients of thalassemia major were taking regular transfusion and iron chelating therapy.

The present study showed that, more than half of the studied children had correct knowledge related to medical treatment of ß-TM. Moreover, the side effect of treatment was unknown for half of the children and adolescents. This could be due to lack of continuous education to children and the nurses did not update their knowledge, as well as, continuing education in children is needed to promote their knowledge and to improve the quality of care. This result was in contrary with that of ***Zaghamir et al.,***

(2019) who carried out a study entitled "Assessment of Thalassemic Children Knowledge about Thalassemia and Iron Chelation Therapy" and reported that most of children and adolescents generally low percentage of satisfactory knowledge regarding treatment of ß-TM and children knowledge very low for the side effect of the drugs. This percentage increased after implementing the educational program. Moreover, this finding goes in line with that of ***Mohamed et al., (2013)*** who carried out a study entitled "Impact of Health Instruction on Improving Knowledge and Practice of Thalassemic Adolescents at Zagazig University Hospital" and found that most of the studied adolescents unknown the side effect of the drugs.

Concerning studied children knowledge about blood transfusion and iron chelation, the findings of the current study revealed that half of them had correct knowledge about the best action when side effect from blood transfusion. In addition, two fifths of them had correct knowledge about foods that increasing the iron in blood. These findings were in accordance with that of ***Chin Kader Maideen & Rashid, (2019)*** who mentioned in a study entitled "Knowledge, Attitude and Practice towards Dietary Iron among Patients with Thalassemia and their Caregivers in Peninsular Malaysia" that majority of the studied subject having the knowledge of foods high in iron because as they learned and acquired some information and advice regarding dietary iron.

As regard to total knowledge about ß-TM of the studied children, the results of the present study illustrated that more than half had unsatisfactory knowledge about ß-TM. This finding comes in line with that of ***Kharyal et al., (20 20*****)** 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 is limited.

Concerning studied children according to their total informational need, the findings of the current study reported that majority of studied children had information needs regards ß-TM. It could be due to there was a positive effect of the educational programme in improving B-thalassemia children's knowledge regarding thalassemia and chelation therapy.

Conclusion:

Based on results of the current study, the present study concluded that more than half of the studied children had unsatisfactory knowledge regarding beta-thalassemia major.

Recommendation:

In view of the study findings, the following recommendations are suggested: providing continuous health education to children with ß-TM regarding their disease. Designing an educational handout about ß-TM and its management plan and allocated for thalassemic children and adolescents. More researchers are needed to explore the factors that affect knowledge deficit by these thalassemic children to generalize the results and be evident to other studies.

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