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Assessment of Knowledge of Adolescents with Thalassemia Major Regarding Iron Chelating Therapy

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Abstract: ***Thalassemia is a genetic blood disease worldwide that is often treated with life-long blood transfusions. Typically every2–4 weeks, this leads to progressive iron overload. Iron chelation therapy is prescribed to manage the transfusional iron overload and attempt to prevent progressive organ failure (heart, endocrine, liver).This study aims to assess the knowledge of adolescents with thalassemiamajor regarding iron chelating therapy.A descriptive design is used in this study that carried out at Hereditary Blood Disease Center in Al- Nasiriya city, a convenience "non-probability sample" of (50) patients of adolescence age (12-18) years old who met the criteria of selection, who attend to hereditary blood disease center, were recruited to participate in this study.Data are collected through the use of a questionnaire tool by direct interview a technique which is used as a means of data collection.The results reveals that the adolescents' knowledge is ranging from poor knowledge 48%, fair 46% and good knowledge 6%.Conclusions of the study indicate that patients need for systematic education to improve knowledge about thalassemia and iron chelating therapy.***

Keywords: Adolescents' Knowledge, Thalassemia Major, Chelating Therapy

1. Introduction

Thalassemia is a genetic, autosomal recessive hemoglobinopathy disease [1]. It is worldwide. It is found in some 60 countries with the highest prevalence in the Mediterranean region, parts of North and West Africa, the Middle East, the Indian subcontinent, Southern Fareast and Southeastern Asia [2,3].

Thalassemia syndromes are characterized by varying degrees of ineffective and increased hemolysis. There are two basic groups of thalassemia disorders: alpha thalassemia and beta thalassemia, varying numbers with each of their particular globin genes mutated [4].

Beta thalassemia, which is a major type of thalassemia, is usually caused by a defect of beta globin protein production. β thalassemia is divided into three categories: thalassemia trait, thalassemia intermedia and thalassemia major (TM). In the first two categories, one of the beta globin genes fails and the quantity of beta globin protein in the cell is reduced by half. In thalassemia major which is also known as "Cooley anemia" the transfusion- dependent clinical phenotype of thalassemia, the absence of β-globin chain production [5,6].

The mainstay of treatment of Thalassemia is blood transfusion combined with iron chelation therapy. While blood transfusion has the advantage of prolonging the patient's life, it is the major cause of iron overloads. About 200 to 250 mg iron that is infused into a thalassemia major patient's body during each blood transfusion that accumulation in many organs resulting in tissues damage [7, 8].

Iron overload occurs when iron intake is increased over a constant period of time, either as an outcome of RBCs transfusions or increased absorption of iron through the gastrointestinal (GI) tract. Both of these occur in thalassemia. When thalassemia major patients receive regularly 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 [9].

The clinical history of thalassemia is changed radically since the emerging of iron chelation therapy prevents the implications of transfusionalhemosiderosis in thalassemia major patients, four decades of experience with deferoxamine visibly has shown the following benefits of iron chelating: Firstly liver iron concentrations can be maintained at normal or mildly elevated levels, secondly can prevent hepatic fibrosis, thirdly can decrease iron induced cardiac disease , fourthly normal growth and sexual development can be achieved and finally long term survival is grandly improved [10,11].

There are three irons chelating drugs currently in use and approved by the U.S. Food and Drug Administration (FDA): deferoxamine, deferiprone, and deferasirox [12].

Thalassemia is a common inherited hematological disorder in Iraq, with an average diffusion of carriers of about 4 % and an estimated 15,000 registered thalassemia major/intermedia patients throughout the country [13].

This study aims to assess the knowledge of adolescents with thalassemia major regarding iron chelating therapy.

2. Methodology

A descriptive study design was used in this study.

Setting of the Study: The present study has been conducted

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on adolescents with thalassemia major at hereditary blood disease Center in Al-Nasiriya city.

Sample of the Study: A convenience "non-probability sample" of (50) patients of adolescence age (12-18) years old who met the criteria of selection, who attend to hereditary blood disease center, were recruited to participate in this study.

Inclusion Criteria

The inclusion criteria include adolescents who are (1) diagnosed with Thalassemia major and receiving iron chelating Therapy, (2) age (12-18) years-old, (3) both genders, (4) are willing to participate in the study, and (5) able to read and write.

The Study Instrument

The study instrument consists of two parts; the first part is related to the participants' demographic which consists of (10) items which include: age, gender, education, residency, age of adolescent at time of diagnosis, age of adolescent at time of starting iron chelating therapy, number of blood transfusion during the last year, number of family members with thalassemia, adolescent's rank among brothers and sisters, and family monthly income. The second part is information related to the thalassemia disease and iron chelating therapy. It consists of (21) multiple choices items (four choices for each question) for assessing adolescents' knowledge about thalassemia and iron chelating therapy. The internal consistency of the questionnaire was (0.95). The data were analyzed by using the statistical package of social science (SPSS) ver. (24.0).

3. Study Results

Table 1: Participants' Demographic Characteristics

Variables	*Groups*	*Frequency*	*Percent*
Age (Years)	12 to less than 15	21	42.0
	15-18	29	58.0
	Total	50	100.0
Gender	Male	25	50.0
	Female	25	50.0
	Total	50	100.0
Education	Able to read and write	9	18.0
	Primary school graduate	17	34.0
	Intermediate school graduate	19	38.0
	Secondary school graduate	5	10.0
	Total	50	100.0
Residency	Urban	37	74.0
	Rural	13	26.0
	Total	50	100.0
Age of Adolescent at Time of Diagnosis	Less than 1 year	33	66.0
	1-<2 years	4	8.0
	2-<3 years	5	10.0
	3-<4 years	4	8.0
	≥ 4 years	4	8.0
	Total	50	100.0
Age of Adolescent at Time of	<5 years old	16	32.0
	≥5 years old	34	68.0
	Total	50	100
Number of Blood Transfusion during the Last Year	> 10 times	2	4.0
	10-20 times	32	64.0
	> 20 times	16	32.0
	Total	50	100.0
Number of Family Members with Thalassemia	1	17	34.0
	2	19	38.0
	3	12	24.0
	4	2	4.0
	Total	50	100
Adolescent's Rank among Family Members with Thalassemia	1st	33	66.0
	2nd	12	24.0
	3rd	5	10.0
	Total	50	100.0
Family Monthly Income (Iraqi Dinar)	< 300 .000	17	34.0
	300.000-600.000	17	34.0
	> 600.000-900.000	10	20.0
	> 900.000	6	12.0
	Total	50	100.0

Results of this table reveal that the most 29(58%) of adolescents in the study sample are within the age group (15-18) years, equally distributed relative to their gender (50%) males and (50%) females, the greater number19 (38%) of them is intermediate school graduates, the greater number of the adolescents are living in urban area and they are accounted 37 (74%) of the sample. Most of the adolescents are diagnosed with thalassemia at the age of less than one year and they are accounted 33(66%) of the sample. Most of the patients start having iron chelating therapy after five years old 34(68%). Regarding to the number of blood transfusion received during the last year, most patients have (10-20) times and they are accounted for 32(64%) of the whole. Concerning to the Number of family members with thalassemia, the greater number of the sample have two family members 19(38%) with thalassemia in the family. Relative to the adolescent rank for family members with thalassemia, 33(66%) of the sample is first one. With regard to the family monthly income, the results present that (34%) have less than 300.000, (34%) have from 300.000–600.000 I.D., (20%) have more than 600.000-900.000 I.D., and (12%) have more than 900.000 I.D.

Table 2: Overall Assessment Adolescents' Knowledge

Level of Assessment	Frequency	Percent
< 50% = Poor (1)	24	48.0
(50- 74) % = Fair (2)	23	46.0
≥75 % = Good (3)	3	6.0
Total	50	100.0
$\bar{x} \mp S.D.$	11.02∓ 3.087	

$\bar{x}$=Arithmetic Mean, S.D.= Standard Deviation,<50% = Poor Knowledge, (50-74%) = Fair Knowledge, ≥75 % = Good Knowledge

Results of this table depict that the adolescents' knowledge about thalassemia and iron chelating therapy is mostly ranging from poor to fair knowledge.

4. Discussion

The study findings indicate that most of the adolescents are diagnosed with thalassemia at the age of less than one year and they are accounted 33(66%) of the sample. Such finding offers empirical evidence that thalassemia can be diagnosed at early age. This is consistent with (Bhatia et. al., 2015) a study on the demographic and morbidity patterns of thalassemia patients registered at a tertiary-care center of

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central India. During the study period, (180) children with thalassemia are identified. Of them, (56.7%) are diagnosed for thalassemia within first 6 months of age [14].

Also, a study that was conducted by Shosha who that examined the beliefs of Jordanian children with thalassemia about using iron chelation therapy. That finding indicates that almost one third of participants were diagnosed before the age of 9 months (37%), and about (63%) were diagnosed at ages between 9 and 12 months [15].

With respect to iron chelating therapy, most of the patients start having iron chelating therapy after five years old. It sounds out of this finding that these adolescents are dependent on such therapy but causes of delay using it may be people do not know the importance of iron chelating therapy and may be lack of treatment in health institutions previouslybecause of its high cost.

Regarding the number of blood transfusion received during the last year, most adolescents have (10-20) times. This finding offers evidence that the adolescents are reliant on blood transfusion as means of treatment.

Concerning the number of family members with thalassemia, the greater number of the sample have two family members with thalassemia in the family. Such finding offers a fact to confirm that thalassemia is inherited health condition. This study is inconsistent with Al-Ali &Faraj study of prevalence of β-thalassemia patients in Missan governorate who demonstrated that more families have one sick child [2].

Relative to the adolescent rank for family members with thalassemia, most are the first one. This finding presents evidence to approve that such health problem can emerge due to marriage of couples of consanguinities and parents' unawareness about disease. The present finding is consistent with a study by Al-Ali &Faraj who demonstrated that affected patients were the highest from first birth order (48.72) than the lowest from fifth birth order (0.51) [2].

With regard to the family monthly income, the finding presents that more than a half have less than 300.000, the same proportion have from 300.000 to 600.000ID, a fifth have 601.00 to 900.000ID, and more than a tenth have more than 900.000 ID. These findings provideevidence that most of the adolescents are living with families at low or middle income and explain that only poor families are more likely to take their children to general hospitals, or may reflect that rich families have enough money to make premarital and prenatal screening tests when they are knowledgeable about it.

With regard to the level of knowledge of adolescent, the finding presents that more number of sample have poor knowledge 48% then 46% have fair knowledge and only 6% have good knowledge (table 2). This refers to lack of awareness of patients regarding thalassemia and its treatment may reflect the lack of educational programs and guidance on this disease in health institutions.

5. Conclusions

Based on the interpretation and discussion of the study findings, the study indicate that patients need for systematic education to improve knowledge about thalassemia and iron chelating therapy.

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