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Factors Affecting Compliance Plan of Thalassemic Children and their Mothers in Outpatient Clinic at Zagazig University Hospitals

Shimmaa Mostafa[1*], Maha Ab Elaziz[2]
1. lecturer of Pediatric Nursing, Faculty of Nursing, Zagazig University, Egypt.
2. lecturer of Community Health Nursing, Faculty of Nursing, Zagazig University, Egypt.
* E-mail of the corresponding author: dr.shimmaa@yahoo.com
Kingdom of Saudi Arabia, Bisha, Riyad Street – Mobil: 00966531646401

Abstract
Children with thalassemia major must be treated with life-long blood transfusions. Evidence suggests that patients with more knowledge/information about their illnesses adhere more readily to treatment schedules. However, there has been little evaluation of disease knowledge and treatment adherence in thalassemia major children. The objectives of the current study were to detect factors affecting the compliance of thalassemic children and their mothers and to identify the relationships between disease knowledge and treatment adherence in thalassemic children. A cross-sectional descriptive design and purposive sampling were used. fifty thalassemia major children (mean age 13.74 years) and their 50 mothers were recruited. structured questionnaire consisting of (A) personal Information; included, Socio-demographic and disease-related data were collected which included age, gender, educational level, (b) An assessment of disease knowledge about TM ; Assessment of disease knowledge of children and their mothers consisted of 20 open questions measuring children's and mothers' knowledge, treatment and compliance of treatment plan about TM. The result of this study revealed that that moderate commitment was apparent in 6% of sample with week knowledge and 60% of sample with moderate knowledge while, strong commitment was found in 26% of sample with moderate knowledge and in 8% of them with week knowledge. This relation was highly statistically significant (p= .000).Based on the findings of the current study, it can be concluded that, The positive association between knowledge about disease and treatment adherence and factors of patients' knowledge indicate the need for systematic education for patients and caregivers to improve their compliance with treatment. The current study recommended that, the improvement of the quality of patient care, reinforcement of medical education and enhanced efforts by clinical staff to provide practical knowledge to children with thalassemia major and their parents is mandatory to significantly improve patient compliance to treatment.
Keywords: thalassemia,compliance.

1. Introduction

Thalassemia is a genetic, autosomal recessive haemoglobinopathic disease, it is found worldwide, but most commonly in the Mediterranean, North Africa and South-East Asia (Faa et al. 2006). Beta thalassemia, a major type of thalassemia, is commonly caused by a defect of beta globin protein production. Beta thalassemia is divided into three categories: thalassemia trait, thalassemia intermedia and thalassemia major (TM). In first two categories, one of the beta globin genes fails and the amount of beta globin protein in the cell is reduced by half. In TM, the transfusion-dependent clinical phenotype of thalassemia, both genes fail and no beta globin protein is produced (Aydinok et al. 2005).

In Egypt carrier rate 5.3 - ≥ 9%, and 1000/1.5 million / year live births born with Thalassemia (total live births 1,936,205 in 2006). Registered cases of homozygous beta thalassemia in big centers of Egypt in 2006 up to Sept 2007 (n=9912) (El-Beshlawy, 2009). Mahmoud, (2009) added that there are 10,000 registered thalassemia cases and more than 20,000 non-registered cases. 95% are beta thalassemia major; 5% are thalassemia intermedia or hemoglobin H disease. Registered cases of homozygous beta thalassemia in Zagazig University Hospital in 2006 up to Sept 2007 (n=710) (El-Beshlawy, 2009).

Worldwide, 15 million people have clinically apparent thalassemic disorders. Reportedly, disorders worldwide, and people who carry thalassemia in India alone number approximately 30 million. These facts confirm that thalassemias are among the most common genetic disorders in humans; they are encountered among all ethnic groups and in almost every country around the world (Yaish, 2009).

TM must be treated by life-long regular blood transfusions (Aessopos et al. 2005). Long-term blood transfusions may generate iatrogenic haemosiderosis, resulting in iron overload. Iron chelation therapy is the mandatory method of preventing organ damage resulting from the accumulation of iron in different tissues (Alymara et al. 2004). Administration of desferrioxamine (DFO) is the primary iron-chelating agent used to treat iron overload (Beshlawy 2005). Iron accumulation can be fatal for TM patients. Most die as a result of complications caused

by ir on accumulation from blood transfusions, with 71% of deaths caused by congestive heart failure owing to iron accumulation in the heart (Cheung et al. 2005). In addition, cardiac disease, pulmonary hypertension (Aessopos et al. 2005), glucose metabolism disorders and gonadal dysfunction can also result from iron overload in TM patients (Platis et al. 2004).

Bone marrow transplantation (BMT) can provide TM patients with an improved chance of survival. Although, some patients have shown great improvement or been cured of TM as a result of BMT treatment, the rigid criteria for screening BMT candidates prevents widespread application of the therapy. Gene therapy is another possible cure for thalassemia (Lin 1997). However, research is still in the experimental stage. The DFO administration is a grueling process.

Clinically, the investigators have observed that some patients did not adhere to regular treatment schedules. Excuses included: 'It is inconvenient', 'I just don't want to do it', 'it hurts' and 'since it leaked out, I don't want to do it again'. Other reasons for unsatisfactory adherence to DFO treatment may be the cost and burden of daily administration (Aydinok et al. 2005). Patients' knowledge about treatment and late effects have been shown to correlate with treatment adherence for many diseases. Studies show that patients with more knowledge of their illnesses are more compliant with treatment schedules: TM (Aydinok et al. 2005); AIDS (Kalichman et al. 2005); and asthma (Schaffer & Tian 2004). Furthermore, several theory-guided intervention studies suggest that the more the information provided to patients, the more were they compliant to treatment: AIDS (Amico et al. 2005), asthma (Schaffer & Tian 2004) and muscular dystrophy. Factors which affect patients' compliance to treatment include individual patient knowledge, attitudes/beliefs, behavioural skills, perceptions of support from significant others, such as caregivers, the therapeutic environment (Amico et al. 2005).

The objectives of the nursing care for children and adolescent with thalassemia start with promote adherence to treatment regimen, as well as support them during illness and distressing treatments, in addition promote patient and family coping such as anticipate older children and adolescents concerns related to appearance and monitor closely for complications of the condition and treatment (Johnson, 2010).

Aims of study:

The current study aims to: (i) detect factors affecting the compliance of thalassemic children and their mothers; (ii) identify the relationships between disease knowledge and treatment adherence in thalassemic children.

Research question:

Is there a relationship between thalassemic children and their mothers knowledge about the disease and compliance to treatment?

Research hypothesis:

- The more the knowledge of the illness, the more adherence with treatment schedules.

Methods

Sample

Cross-sectional descriptive design and purposive sampling were used. Eligible patients were chosen from TM patients who attended the out patient Pediatric Hematology clinic at Zagazig University Hospital in Zagazig City. Inclusion criteria were children with TM who: (i) were able to communicate with the researcher (ii) had no severe mental illness diagnosed by psychologists or psychiatrists; (iii) were 10 years or older; and (iv) gave informed consent. Fifty patients were potentially eligible. Patients of 10 years and older were included because they had already been treated for TM for several years and were expected to have TM-related knowledge to take care of themselves. After receiving informed consent, patients were individually administered a questionnaire by a researcher. Data from 50 patients and their mothers were used for analyses.

Measure (tools):

Data were collected by structured questionnaire consisting of:

(a) A personal Information; included, Socio-demographic and disease-related data were collected which included age, gender, educational level, employment, monthly household income, haemoglobin level, duration and frequency of blood transfusions, duration and frequency of DFO infusions and importance and meaning of commitment to treatment plan.

(b) An assessment of disease knowledge about TM ; Assessment of disease knowledge of children and their mothers consisted of 20 open questions measuring children's and mothers' knowledge, treatment and compliance of treatment plan about TM. Each item was scored using 0 , 1 , 2 (0 = incorrect/do not know; 1 = correct but incomplete, 2= correct and complete answer). A higher score represented a higher level of knowledge about TM and its treatment plan and importance of compliance and reasons for in commitment. The questionnaire was developed based on a literature review, personal clinical observations and suggestions from two paediatric haematologists. Two paediatric haematologists and two senior paediatric nurses examined the instrument for the appropriateness, representativeness and relevance. Face validity and content validity were used to

evaluate the questionnaire. Comments from the experts indicated that it was an appropriate measure for a construct of TM-related knowledge. Five children and their mothers in this study, were invited to pilot test the questionnaire. The pilot-study patients reported that the questionnaire was easy to understand.

Procedure

Questionnaires were administered to the target sample by a researcher as patients were receiving blood transfusions. Patients could ask questions during the process. Accompanying parents were told not to help with answers. Parents could sit beside patients during data collection and filled out the same questionnaire as the patients. mothers and children were informed that no individual's identity would be disclosed and that they could refuse to answer any questions or to terminate the survey at any time. Informed consent was obtained prior to filling out the survey. The survey took about 10–15 minutes to complete.

Analyses

Data were analyzed using SPSS for Windows. Descriptive statistical methods were used. Pearson correlation was used to examine the relationships between patients' knowledge and treatment adherence, some demographic and disease variables and parents' knowledge. Perspectives of patients and family carers knowledge and adherence.

Table (1): Socio-demographic characteristics of TM children and their mothers.

Characteristics	N (n=50)	(%)
Age Mean ± SD	13.74±2.107	
Sex		
Male	33	66.0
Female	17	34.0
Birth order		
First	16	32.0
Second and third	26	52.0
Forth or more	8	16.0
Residence		
Urban	20	40.0
Rural	30	60.0
Mothers age in years		
20-30	20	40.0
30-40	12	24.0
≥40	18	36.0
Mother Education		
Illiterate	19	38.0
Read & write	6	12.0
Secondary	23	46.0
University	2	4.0
Mother Occupation		
Working	9	18.0
Not working	41	82.0
Family monthly income		
Sufficient and save	6	12.0
Sufficient	23	46.0
Insufficient	21	42.0

Table (2): Medical history and physical activity of TM children

Medical History/ physical Activity	N (n= 50)	(%)
Family history of thalassemia		
- Yes	30	60.0
- No	20	40.0
Consanguinity relation with thalassemic child		
- First degree	18	36.0
- Second degree	12	24.0
- Other	20	40.0
Does the child practice sport or activity?		
▪ **Yes**	22	44.0
▪ **No**	28	56.0
Type of sport to be practiced		
▪ **Walking**	12	32.0
▪ **Running**	2	4.0
▪ **Swimming**	1	2.0
▪ **Football**	7	16.0
▪ **None**	28	56.0
Is the child feel tired after exerting effort?		
▪ **Yes**	44	88.0
▪ **No**	6	12.0
Manifestation of child's tiredness:		
Headache	2	4.0
Nausea	8	16.0
Sweat	9	18.0
Dyspnea	25	50.0
None	6	12.0

Table (3): knowledge of the TM children and their mothers about the disease

Factors	Mothers (n= 50)		TM children (n= 50)	
	No	%	No	%
Definition of thalassemis				
- Don't know or incorrect	32	64.0	38	76.0
- Correct but incomplete	17	34.0	12	24.0
- Correct and complete	1	2.0	0	0.0
Prevalence of thalassemia				
- Don't know or incorrect	23	46.0	30	60.0
- Correct but incomplete	0	0.0	0	0.0
- Correct and complete	27	54.0	20	40.0
Risk factors of disease				
- Don't know or incorrect	21	42.0	34	68.0
- Correct but incomplete	28	56.0	15	30.0
- Correct and complete	1	2.0	1	2.0
Diseases' symptoms				
- Don't know or incorrect	9	18.0	22	44.0
- Correct but incomplete	25	50.0	17	34.0
- Correct and complete	16	32.0	11	22.0
Complications of disease				
- Don't know or incorrect	12	24.0	30	60.0
- Correct but incomplete	21	42.0	10	20.0
- Correct and complete	17	34.0	10	20.0
Effect of iron precipitation				
- Don't know or incorrect	29	58.0	34	68.0
- Correct but incomplete	20	40.0	15	30.0
- Correct and complete	1	2.0	1	2.0

Table (4): Factors related knowledge of TM children and their mothers about treatment plan

Factors	Mothers (n= 50)		TM Children (n= 50)	
	No	%	No	%
Importance of treatment with Desferal				
- Don't know or incorrect	25	50.0	34	68.0
- Correct but incomplete	25	50.0	16	32.0
- Correct and complete	0	0.0	0	0.0
How many times desferal should be taken?				
- Don't know or incorrect	21	42.0	27	54.0
- Correct but incomplete	25	50.0	20	40.0
- Correct and complete	4	8.0	3	6.0
Side effect of desferal				
- Don't know or incorrect	31	62.0	35	70.0
- Correct but incomplete	19	38.0	15	30.0
- Correct and complete	0	0.0	0	0.0
Precautions to avoid desferal side effects				
- Don't know or incorrect	32	64.0	46	92.0
- Correct but incomplete	18	36.0	4	8.0
- Correct and complete	0	0.0	0	0.0
Hb level required for blood transfusion				
- Don't know or incorrect	28	56.0	40	80.0
- Correct but incomplete	22	44.0	10	20.0
- Correct and complete	0	0.0	0	0.0
Importance of blood transfusion				
- Don't know or incorrect	25	50.0	34	68.0
- Correct but incomplete	25	50.0	16	32.0
- Correct and complete	0	0.0	0	0.0
Complication of stopping blood transfusion				
- Don't know or incorrect	27	54.0	37	74.0
- Correct but incomplete	23	46.0	13	26.0
- Correct and complete	0	0.0	0	0.0
Complication of recurrent blood transfusion				
- Don't know or incorrect	13	26.0	25	50.0
- Correct but incomplete	34	68.0	24	48.0
- Correct and complete	3	6.0	1	2.0
Causes of sever complications				
- Don't know or incorrect	12	24.0	32	64.0
- Correct but incomplete	36	72.0	17	34.0
- Correct and complete	2	4.0	1	2.0
Meaning of commitment to treatment plan				
- Don't know or incorrect	13	26.0	28	56.0
- Correct but incomplete	37	74.0	22	44.0
- Correct and complete	0	0.0	0	0.0
Importance of commitment to treatment plan				
- Don't know or incorrect	13	26.0	25	50.0
- Correct but incomplete	37	74.0	25	50.0
- Correct and complete	0	0.0	0	0.0

Table (5): Relationship between importance of commitment to treatment plan and sociodemographic characteristics of TM children and their mothers.

Sociodemographic Characteristics	Mother's knowledge about the importance of commitment to treatment plan (n = 50)				TM children knowledge about the importance of commitment to treatment plan (n = 50)			
	Don't know or incorrect		Correct but incomplete		Don't know or incorrect		Correct but incomplete	
	No.	%	No.	%	No.	%	No.	%
Mother education								
Illiterate	8	42.1	11	57.9	8	32.0	11	44.0
Read & write	3	50.0	3	50.0	4	16.0	2	8.0
Secondary	2	8.7	21	91.3	11	47.8	12	52.1
University	0	0.0	2	100.0	2	100.0	0	0.0
(p value)	.045				.139			
Mother occupation								
Working	3	33.3	6	66.6	6	66.6	3	33.3
Not working	10	24.4	31	75.6	19	46.3	22	53.7
(p value)	.580				.269			
Residence								
Urban	11	36.7	19	63.3	15	50.0	15	50.0
Rural	2	10.0	18	90.0	10	50.0	10	50.0
(p value)	.035				1.000			
Family monthly income								
Sufficient and save	0	0.0	6	100.0	5	83.3	1	16.7
Sufficient	9	39.1	14	60.9	13	65.5	10	43.5
Insufficient	4	19.1	17	80.9	7	33.3	14	66.7
(p value)	.096				.067			

Figure (6):Relationship between disease related knowledge of TM children and their mothers and their commitment to treatment plan.

Commitment to treatment plan	Disease related knowledge of studied children and their mothers				Total	
	Week Knowledge		Moderate knowledge			
	No.	%	No.	%	No.	%
Moderate Commitment	3	6.0	30	60.0	33	66.0
Strong Commitment	4	8.0	13	26.0	17	34.0
(p value)	.000					

Results

Sample sociodemographic characteristics of both thalassemic patients' and mothers' knowledge, treatment adherence, the relationships between knowledge and treatment adherence in patients; are as follows.

Regarding the thalassemic children, fifty thalassemic child, aged 10–18 years, (mean±SD = 13.74±2.107), were included. Most patients were male (66%), more than half of TM children were from rural area. Concerning TM children mother's, fifty mothers were included (40%) of them aged 20-30 years while mothers aged 40 years or more were (36.0%). Thirty eight percent of mothers were illiterate while (40%) percent of them were Diplomat education and only 4.0% were university education. Majority of mothers (82.0%) were not working and (42.0%) of them had insufficient monthly income and only 12.0% of them had sufficient income and could save based on the self-report of mothers (table 1).

About family history of thalassemia (36%) of TM patients had consanguinity relation with relative in the first degree who had the disease (table 2) Regarding the sports which practiced by TM patients , it was found that

(56%) of TM patients practiced sports and walking was the most sport type practiced by 32% of TM patients. The majority of TM patients (88%) felt tired after exerting effort. Half of TM patients (50%) had dyspnea and 18 % of them had sweat as manifestations of tiredness after exerting effort.

According to the disease knowledge in both TM patients and their mothers (table 3), about two thirds of mothers (64%) and (76%) of TM patient didn't know or gave incorrect answer about the definition of thalassemia. More than half of mothers (54%), and more than one third (40%) of TM patients answered correct and complete about the item of thalassemia prevalence while about one half (46%), and about two thirds (60%) of mothers and TM patients respectively didn't know or gave incorrect answer. Concerning the disease risk factors item, the majority of mothers (56%) and nearly one third (30%) of TM patients didn't know or answered incorrectly. Forty two percent of mothers and twenty percent of TM patients answered correct but incomplete about complications of disease item, while (34%), and (20%) of mothers and TM patients respectively their answers were complete and correct. Regarding mothers and TM patient knowledge about the effect of iron precipitation, only 2% answered correctly and completely.

Concerning mothers and TM patient's knowledge about treatment plan (table 4), half of mothers (50%) and majority of TM patients (68%) didn't know or gave incorrect answer, while non of them answered in correct or complete about the importance of treatment with desferal. Half of mothers (50%) and less than half of TM patients (40%) gave correct but incomplete answer about how many times Deciferal should be infused. Regarding their knowledge about side effect of desferal majority of mothers and TM patients (62%), (70%) respectively didn't know or answered incorrectly, on the other hand non of them gave correct or complete answer. None of the mothers or TM patients answered correctly and completely about precautions to avoid desferal side effects, while only 8% of TM patients answered correctly but incomplete. Concerning the item of required Hb level for blood transfusion, more than half of mothers (56%) and majority of patients (80%) didn't know or gave incorrect answer, while none of them answered correctly or completely. Concerning the importance of blood transfusion, half of mothers (50%) and more than two thirds (68%) of TM patients answered incorrectly, 50% of mothers and 32% of TM patients answered correctly but not complete about these item. As regard the item of stopping blood transfusion complications knowledge, more than half of mothers (54%) and about three fourths of TM patients (74%) answered incorrectly or didn't know the answer, while none of them know the correct answer. Sixty eight percentage and forty eight percentage of mothers and TM patients respectively, gave correct but incomplete answer about the item of complications of repeated blood transfusion.

When mothers and TM patients were asked about the importance of commitment to treatment plan (table 4), about one quarter (26%) of mothers and one half (50%) of TM patients answered incorrectly or didn't know, and (74%), (50%) of mothers and TM patients respectively answered correctly but incomplete and none of them know the correct and complete answer.

Relationships between TM patients and their mothers' knowledge about the importance of compliance of treatment plan, it was found that there was statistical significant difference between mothers' knowledge about importance of commitment to treatment plan and their educational level (p=0 .04*) , and residence (p=0.03*) (table 5).

According to the relation between the sample knowledge and their compliance to treatment plan (table 6), it was found that , moderate commitment was apparent in 6% of sample with week knowledge and 60% of sample with moderate knowledge while, strong commitment was found in 8% of sample with week knowledge and in 26% of them with moderate knowledge. This relation was highly statistically significant (p= .000).

Discussion:

The present study showed that more than half of the studied children were males. This finding is in consistent with the findings of Elsaid (2009) who done his study at Zagazig university Hospitals, Zagazig, Egypt and Salama et al (2006) who done his study at Mansoura University Children's Hospital, Mansoura, Egypt. The present study is also supported by Shaligram et al (2007) who done his study in Shiraz city. Similar to Shaligram et al (2007), the present study showed that about two thirds of children were from rural area. On other hand, the current study is in contrast with Gharaibeh et al (2009) who done his study at the National Thalassemic Center in Damascus, Syria. Gharaibeh study illustrated that about three quadrants were from urban. This reflects that thalassemia may be presented in urban as in rural when premarital screening and genetic counseling is neglected. In addition, the data was collected from Pediatric Hematology Outpatient Clinic at Zagazig University Hospital that represents the only center serving thalassemic patients all over Sharkia Governorate for free and attracts patients from all villages of the governorate.

The current study found that most families reported insufficient and sufficient family income. This finding goes in line with the findings of Gharaibeh et al (2009). This may 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.

Shaligram et al (2007) noticed that about half of the cases were outcomes of first-or second-cousin marriages.

Also, Naseri (1997) revealed in his study that 66% of thalassemic children had consanguineous parents. This goes in line with the present study which revealed that more than half of parents of the studied children were relatives. This may be due to strong family relationships in Egypt, especially those who live in rural areas and that the closer the relation between the parents, the greater the risk that many children might be born with a hereditary disorder such as thalassemia.

Tadmouri et al. (2012) stated that Arab populations have a long tradition of consanguinity, due to sociocultural factors such as maintenance of family structure and property, and ease of marital arrangements. Many Arab countries display some of the highest rates of consanguineous marriages in the world, specifically first cousin marriages. The main impact of consanguinity, however, is an increase in the rate of homozygotes for autosomal recessive genetic disorders such as thalassemia. In addition, educational programs about genetic counseling are still neglected.

Eleftheriou (2007) clarifies that about 2-3 children in every 1,000 born to unrelated parents have a recessive disorder, while from 2-20 children in 1,000 born to related parents have a recessive disorder, depending on how closely the parents are related.

Gharaibeh et al (2009) found that more than two thirds of children had sick relatives with thalassemia. This goes in line with the present study where more than half of children had family history of thalassemia. This may be the result of increased consanguineous marriages.

Many factors can affect daily living physical activities in thalassemic patients such as chronic hypoxia that results from progressive anemia and leads to decreased exercise tolerance (Hockenberry & Wilson, 2009). The results of the present study showed that less than half (44%) of the thalassemic patients practiced some kind of sports or activities and 16% those played football. The male predominance among the studied sample played a role in this result as well as the predominance of this age group, since they tempted to risk-taking behaviors, especially males.

The current study reveled that about one third of thalassemic patients had no problem with walking, this is goes in line with Shaligram et al (2007) who found that the majority of children had no problems with mobility.On the other hand, more than half (56%) of the thalassemic patients did not practice any kind of sports or activities. As a result of fear of feeling tired and exhausted after doing any effort as the main cause of not practicing sports or activities. This is goes in line with Khurana et al. (2006) who studied the psychosocial life aspects of Indian adolescents suffering from β-thalassemia major and reported that over two thirds of the adolescents were unable to engage in outdoor play at the same level as their peers, because of physical weakness related to their disease. Indulgence in sport activities was therefore limited. Also, Caro (2002) who found that, less than one quadrant of conventionally treated thalassemia major patients had their activities very often stopped due to thalassemia, its complications or desferrioxamine treatment, and 20% had their physical activities limited at least a bit. This may be due to the regular period of mild anemia before the scheduled of transfusion which might limit their exercise capacity as thalassemia leads to low hemoglobin level resulting in fatigue and general weakness (Cheuk et al, 2008).

Regarding to thalassemic patients and their mothers knowledge about definition of thalassemia, about two thirds (76%) and (76%) of them respectively were not able to define thalassemia correctly or gave incomplete answer. This is may return to lack of health education about the disease itself and that the health team either doctors or nurses tend to give more health instructions about the care practices than the disease. El-Awany (2002) and Hassan (2009) were in agreement with these results. They noted that most of the children had wrong information regarding definition of thalassemia before implementation of the educational programs, and there was a significant improvement of knowledge after implementation of the educational programs in both studies.

The results of the present study revealed that only twenty percentage of thalassemic patients and thirty four percentage of their mothers answered complete and correctly. Yang et al. (2005) supported these findings and reported that, the worst knowledge score in his study was the knowledge of complications. The bad knowledge about complications in the present study may be explained through the fact that, many patients avoid receiving complications-related information owing to distress about an uncertain future.

In a study conducted to explore the relationships among illness knowledge, social support and self-care behavior in adolescents with β-thalassemia major in Taiwan, Yang et al. (2005) reported that treatment knowledge was the best among the scores of illness knowledge. This was not the case in the present study where more than half of mothers and majority the thalassemic patients were not able to give correct answer or don't know importance of desferal to treat thalassemia.

Hashem (2006) noticed that, thalassemic children had average level of understanding the importance of blood transfusion therapy before educational program implementation. After their attendance and participation in the

program, they gained more knowledge about the importance of blood transfusion therapy. In the current study, regarding importance of blood transfusion half of mothers (50%) and about one third of thalassemic patients (32%) answered correctly but incomplete as regard the importance of blood transfusion to improves general health, increases activity, provides energy, prevents blood destruction and prevents fatigue.
The previously mentioned incorrect answers originated from the fact that the some of the thalassemic patients did not have blood transfusion unless their hemoglobin level become very low and start to suffer from hypoxic manifestations [headache, precordial pain, bone pain, decreased exercise tolerance, listlessness and anorexia (Hockenberry & Wilson, 2009) and these manifestations improved significantly after the blood transfusion and this was clear in their understanding of the blood transfusion importance.

In the current study, the hemoglobin (Hb) level that required for blood transfusion in thalassemic patients was not known to more than half of the mothers and majority of thalassemic patients. This is contradicted with Lee et al. (2008) who reported that more than three quarters of his studied patients were able to determine the mean Hb level (9-10.5 g/dl) that required in thalassemic patients for transfusion. Lack of knowledge observed among thalassemic patients in the current study may be due to two reasons (i) inadequate explanation from health-care providers and/or (ii) most of the patients and their mothers believed that investigations, Hb level and treatment issues are the responsibility of the physician and they do not need to know a lot about it (iii) poor socioeconomic status of most of the studied patients and getting sick of so many needles they had to have during treatment process. Porter, 2011 stated that monitoring pre-transfusion Hb level is very important to prevent complications.

Concerning the studied thalassemic patients and their mothers knowledge about side effects of desferal, it was found that, the side effects of desferal were not known to more than half of the mothers (62%) and majority of the thalassemic patients (70%). This is may be due to the belief that, if the patient knows the possible side effects of the drug they will be afraid of taking it and consequently will not comply with it. So, health care providers usually tend to avoid the discussion of medication's side effects with the patients. Moreover, the pamphlets of all iron chelating agents were written in English language, that the studied patients cannot read or understand.
Same knowledge deficit regarding chelation therapy was reported by EL-Awany (2002) who found that, thalassemic children and their mothers lacked the essential knowledge in this area before implementation of the educational program. Nevertheless, after implementation of the health instructions, thalassemic adolescents' knowledge improved significantly regarding side effects of desferal, kelfer, ferriprox and exjade.

As regards the thalassemic patients and their mothers knowledge about importance of compliance with treatment plan, majority of mothers (74%) and half of thalassemic patients (50%) answered correctly but incomplete about, compliance with treatment decrease iron overload. This is congruent with Lee et al. (2008) who reported that, the majority of his studied patients stated that, importance of compliance with iron chelators is to get rid of excess iron and to avoid iron sedimentation. This may be due to, the stress of physicians' instructions on the importance of iron chelators in reducing iron overload.

This study shows that TM patients and their mothers' knowledge about the importance of compliance of treatment plan and mother's education was statistically significant relation (p=.045), mothers' knowledge about TM is dependent on mother's educational level and that is the most significant variable influencing patients' knowledge. This supports the literature: In a survey of 38 patients with TM and their mothers, Aydinok et al. (2005) reported that mothers' educational and psychological burden correlated with the patients' mental status and that the more disease awareness possessed by both mothers and patients, the more compliant patients were to related treatment. It is not surprising that the effect of mothers' knowledge on patients' knowledge is tremendously significant as mothers are the main caregivers. Teaching and research should therefore pay attention to educating patients' mothers about TM in their children.

There was no significant relationships between TM patients and their mothers' knowledge about the importance of compliance of treatment plan and family monthly income (p=.096). In the past, funding was a serious issue during the TM treatment. This expenditure was a huge burden for a middle-class family. This cost may have deterred patients and their families from taking DFO infusions on schedule. They may have instead chosen to inject DFO only when serum ferritin accumulated and exceeded the normal range. After implementation of the Egyptian National Health Insurance .Economic limitation as a factor is expected to have been reduced. Moreover, the efforts of several patient and social support associations, might have helped to improve patients' adherence to blood transfusions and DFO infusions.

The present study found that moderate commitment was found in 6% of studied children and their mothers who had week knowledge and 60% of them with moderate knowledge while, strong commitment was apparent

in 8% of the studied sample with week knowledge and in 26% of them with moderate knowledge. This finding gives an objective indicator to assess the level of disease knowledge in TM patients and their mothers. Patients with poor compliance to treatment plan may have less TM knowledge.

This finding is supported by Elsaid (2009) who done his study at Zagazig university Hospitals, Zagazig, Egypt and he found the most of his studied sample were not complaint with iron-chelation therapy. Poor knowledge of thalassemic patients and their mothers regarding the disease, and the treatment plan, is a multifaceted issue. As, it may be owing to; excessive anxiety, fear of condition, illiteracy, impaired communication, lack of prior teaching, misinterpretation of information and unwillingness to learn. In addition, lack of adequate guidance by the health care providers and lack of effective communication between the nursing staff and the patients also play a role in this knowledge deficit. In the study setting, (Hematology Outpatient Clinic) there is a shortage in the nursing staff number especially the highly qualified nurses, who supposed to be able to give adequate explanation about the disease and its care aspects. This shortage in the nursing staff number represents another important issue in this problem.

Conclusion

Based upon the findings of the current study, it can be concluded that, positive association between knowledge about disease and treatment adherence and factors of patients' knowledge indicate the need for systematic education for patients and caregivers to improve their compliance with treatment.

Recommendations:

- Improvement of the quality of patient care, reinforcement of medical education and enhanced efforts by clinical staff to provide practical knowledge to children with thalassemia major and their parents is mandatory to significantly improve patient compliance to treatment.
- Age-appropriate cartoon illustrations for children to provide detailed scientific information about the disease and how to survive with it.
- 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.
- Mass media must provide population with genetic counseling about thalassemia and other inherited diseases and discourage consanguineous marriage.
- Further researches are needed to investigate factors affecting compliance with treatment regimen.

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