Relationship between Knowledge of Patients with Thalassemia and their Quality of Life

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Abstract

Background: Thalassemia is one of the most common genetic disorders worldwide that associated with defective hemoglobin-chain synthesis. Clinical manifestations are diverse, ranging from asymptomatic hypochromia and microcytosis to profound anemia. **Aim of study:** This study aimed to assess the relationship between knowledge of patients with thalassemia and their quality of life through: (1) Assessment of level of knowledge for adult patients suffering from thalassemia, (2) Assessment of quality of life among adult patients suffering from thalassemia. **Research design:** A descriptive exploratory research design was utilized to achieve the aim of the present study. **Setting:** The current study was carried out in general medicine department at El Fayoum university hospital. **Subject:** A purposive sample of 80 adult patients with thalassemia was included in this study. **Tools of data collection**: First tool: Patient interview questionnaire. Second tool: World Health Organization Quality of Life-BREF (WHOQOL-BREF) questionnaire. **Results:** The results of this study revealed that more than two thirds of patients had satisfactory level of knowledge regarding thalassemia, and more than three quarters of patients had poor quality of life. **Conclusion:** It was concluded that there was no statistical significance relation between total level of knowledge of patients with thalassemia and their quality of life. **Recommendations:** Regular follow up for patients with thalassemia to evaluate their health conditions and to detect complications early. Further studies are needed to assess factors affecting quality of life of patient with thalassemia.

Keywords: Knowledge - Quality of Life - Thalassemia - Patients.

Introduction:

The term ‘thalassemia’ refers to a genetic blood disease. Thalassemia is the most prevalent inherited hemo-globinopathy that result from a decreased synthesis of alpha or beta chains of hemoglobin (Hb) **(Bhurani et al., 2021)**.

Thalassemia is the most common hereditary anemia in the world with approximately 300 million carriers. This hemoglobinopathy is more prevalent in East Mediterranean region, Southeast Asia and India. Thalassemia also is a major health problem in the northern and southern provinces of Iran **(Jafari, Davoudi, A.Pour & Mohtasham, 2022).**

It is the commonest type of chronic hemolytic anemia in Egypt and the most frequent hemoglobin disorder in the country. The carrier rate of this disease varies between 5.3 and 9% **(Ragb, Elrahim, El Fotoh & Ibrahim, 2021)**. It is estimated that the average annual incidence is one in 100,000 **(Saad, Bahgat, El Dewy & Galal, 2021).**

Patients with thalassemia are marked with symptoms such as chronic and severe anemia, growth retardation, enlargement of the spleen and liver, bone disorders, especially visible changes in the craniofacial skeleton, heart failure, pancreatic disorders, and hepatitis B, C, D and E. The treatment basis for these patients is a repeated blood transfusion that can treat anemia. However, it causes iron overload in vital organs, including the heart, pancreas and

liver, which disrupts the function of these organs. So patient needed to iron chelation therapy **(Ghorbanpoor, Mirzaie, Mirhaghjou & Roshan, 2020).**

Knowledge of the disease involves a range of beliefs based on the information about various aspects of the disease that the patient has collected over life, both before and after the diagnosis. The knowledge usually pertains to the causes of the disease and exacerbating factors, identification of symptoms, and available methods of treatment and consequences **(Man, Maideen & Rashid, 2019).** So, raising awareness about the disease is an essential component of successful thalassaemia prevention **(Hossain et al., 2020)**.

The quality of life is defined as the characteristics that people require to maintain their abilities to conduct valuable life activities and the chronic diseases has negative effects on the health and quality of life. Today, the quality of life is considered an important issue and consequence of health in treating and taking care of chronic patients. As chronic disease, survival and Quality of Life (QoL) of the patients are of the most importance and are influenced by psychological and social constraints **(Senthil, Mahmood & Grainger, 2022)**.

Significance of the study:

Thalassemia is a chronic disease that presents a diverse range of serious clinical and psychological challenges. These challenges not only do effect on the patients' physical, emotional and social functioning but also their quality of life and relationship with their families **(Maheri, Rohban, Sadeghi & Joveini, 2020)**.

In spite of the treatment measures that increase the survival of these patients, its clinical signs adversely affect the physical and mental health of the patients and their family. These physical and psychological problems lead to disappointment, decreased function, and ultimately reduced the quality of life. Not only factors such as insufficient knowledge, course duration of the disease and treatment, hospitalization, repeated laboratory tests for monitoring therapy and for early detection of any side-effects or complications and increased medical costs, mental status, and social problems cause stress in patients with chronic disease and their family, but also its effects on the patient's quality of life. So, when there are enough definite knowledge with those pre-existing factors, patient's quality of life likely to be improved **(Ahmadi et al, 2020)**.

Aim of the study:

This study aimed to assess the relationship between knowledge of patients with thalassemia and their quality of life through:

1- Assessment of level of knowledge for adult patient suffering from thalassemia.

2- Assessment of quality of life among adult patients suffering from thalassemia.

Research questions:

1-What is the patients' level of knowledge regarding thalassemia?

2-What is the level of quality of life for patients with thalassemia?

3-What is the relationship between patient's knowledge and their quality of life regarding thalassemia?

Subjects and methods:

Research design:

A descriptive exploratory research design was utilized to achieve the aim of the present study.

Setting:

The current study was carried out in general medicine department at El Fayoum university hospital.

Subject:

A purposive sample of 80 adult patients with thalassemia was included in this study based on the following criteria:

Inclusion criteria: Adult patient from both genders who are diagnosed with thalassemia, able to communicate verbally and willing to participate in the study.

Exclusion criteria: psychiatric patients, pediatric patients and patients suffering from other types of anemia.

Tools for data collection:

Two tools were used in this study:

Frist tool: Patient interview questionnaire:

This tool was developed by the researcher in an Arabic language based on the related literatures **(Haq et al., 2017; Olwi, Merdad & Ramadan, 2018; Man et al., 2019).** It was used to assess knowledge of patients suffering from thalassemia. It consists of 66 questions in form of closed, open ended and multiple-choice questions. The tool consisted of three parts:

Part I: Patients' demographic data:

This part aimed to assess patients' demographic data such as patients' age, gender, educational level, marital status, occupation, type of occupation, residence, living condition and income. It contains nine questions in form of multiple-choice questions and closed ended questions.

Part II: Patients' medical history:

This part aimed to assess patients' medical history that includes information about the present history, duration of the disease, past history, previous hospitalization and family history of thalassemia. It contains 12 questions in form of closed, open ended and multiple-choice questions.

Part III: Patients' knowledge assessment regarding thalassemia:

This part aimed to assess patients' knowledge regarding thalassemia. It includes questions about definition, causes, signs and symptoms, diagnostic test, prognosis, complications, treatment, iron therapy, diet and activity. It consisted of 45 questions in form of multiple-choice questions.

❖ **Scoring system:**

The scoring system of this part as the following:

- The tool consisted of 45 questions.
- The response for each question was by choosing the correct answer.
- The correct answer was given one grade.
- The incorrect answer was given zero.
- The total grade for this part was 45 grades.
- The total score of all patients were calculated as number and percentage.

The score was classified into 2 categories:

- Satisfactory level of knowledge if scores ≥60% (27-45 degree)
- Unsatisfactory level of knowledge if scores <60% (<27degree)

Second tool: World Health Organization Quality of Life-BREF (WHOQOL-BREF) questionnaire:

The WHOQOL-BREF was developed by **GENEVA, 1996.** It was used to assess quality of life for patient with thalassemia. It was translated into an Arabic language and back translated into English. The tool included (26) questions.

❖ **Scoring system:**

This tool is divided into 4 domains: physical domain (7 items), psychological domain (6 items), social domain (3 items), and environmental domain (8 items), as well as the overall quality of life and general health (2 items). Scoring of each item is rated on a five-point likert scale that ranged from 1 to 5 where 1 is the lowest score and 5 are the highest score, (1=1) (2=2) (3=3) (4=4) (5=5). There are three negative phrased items (Q3, Q4, and Q26) which is reversely scored. (1=5) (2=4) (3=3) (4=2) (5=1). This transforms negatively framed questions to positively framed questions. The total grade for this questionnaire is 130 grades that represent 100%.

The score were categorized into:

- Good quality of life: ≥60% (78-130)
- Poor quality of life: <60% (<78)

Content validity:

The validity of the developed tools was tested by a panel of seven experts from medical surgical nursing department, faculty of nursing, Ain Shams University. The panel of experts was from different academic categories (five professors and two assistant

professors). The experts reviewed the tools for clarity, relevance, comprehensiveness, understanding, applicability and easiness for administration. According to their suggestions, some modifications were done (where some items and questions were omitted and others were added based on recommendation from the experts).

Reliability:

Testing reliability of the developed tool was done by cronbach's alpha test. It was 0.889 for total knowledge assessment questionnaire. Testing reliability of the standerd tool was done by Cronbach's alpha coefcient test. It was 0.70 for WHOQoL-BREF questionnaire **(Floris et al 2018)**.

Ethical consideration:

The ethical research consideration included the following:

- The research approval was obtained from the ethical research committee at Faculty of Nursing Ain Shams University before initiating the study work.
- The researcher clarified the objectives and aim of the study to patients included in the study.
- Oral consent was obtained from patients before participation in the study.
- The researcher assured maintaining anonymity and confidentiality of subjects' data.
- Patients were informed that they are allowed to withdraw from the study at any time without giving any reasons and without penalties.

Pilot study:

A pilot study was carried out on 10% (8) of the patients under study to evaluate the applicability, clarity, efficiency and time needed to fill in each tool. Also, it help to find the possible obstacles and problems that might face the researcher and interfere with data collection. The necessary modifications were done, where some items and questions were omitted and others were added based on the result of the pilot study. The eight patients who included in the pilot study were excluded from the study.

Field work:

- The collection of data of the current study continues for six months from September 2021 until the end of February 2022.
- An approval was obtained from hospital directors and nursing directors of general medicine department in El-Fayoum university hospital.
- The researcher visited the study setting 2 days (Monday and Thursday) per week from 9 am to 3 pm to gather data using the previously mentioned tools.
- The purpose of the study was explained to patient under study prior to any data collection.
- Oral consent was obtained from patients after explaining the aims of the study.
- The researcher started the interview with each patient individually using the data collection tools.
- The time consumed to fillin the tool by the researcher: 20 minutes for Patient interview questionnaire and 10 minutes for WHOQoL-BREF questionnaire.

IV. Statistical design:

Data collected and coded to facilitate data manipulation and double entered into Microsoft Access and data analysis performed using the Statistical Package of Social Science (SPSS) software version 22 in windows 7. Independent samples t test and One-way ANOVA test was used to find the association between variables of quantitative_data. The chi-square ($\chi 2$) test was used to find the association between variables of qualitative data. Cronbach's alpha test of reliability, The P-value< 0.05 considered as statistical significant

Results:

Table (1): Showed that 52.5% of the studied patients their age ranged from 18-24 years old. As regard to gender, the table revealed that 62.5% of patients were males and 37.5% were females. Regarding to marital status, it was revealed that 72.5% of patients were single and 22.5% were married. Regarding to educational level, the result showed that 25% of patients can't read and write 45% had primary education. In relation to occupation, it shows that 72.5% of patients don't work and

25% of them work as part time. Concerning residence, 72.5% of patients under the study are living in rural inhabitation and 27.5% of them from urban inhabitation. This table also shows that 72.5% of patients don't have enough income as they reported and 27.5% of them had enough income. In relation to living condition, the result shows that 100% of patients are living with their families.

Table (2): Illustrated that the higher satisfactory knowledge level among patients with thalassemia was 4.6± 1.1 about blood transfusion therapy, followed by 4.1± 1.5 about healthy diet, then 3.9±0.72 for thalassemia diagnosis methods, 3.8±0.52 for physical activity, 3.4±1.3 for complications of thalassemia. While the lowest knowledge score for patients with thalassemia was 1.7±1.2 for definition, cause and types of thalassemia, 2.7±1.4 for signs and symptoms, 3.1±0.84 for treatment of thalassemia, 3.2±2.1 for chelating therapy. The result revealed that total knowledge score among patients with thalassemia was 30.4±7.5.

Table (3): Showed that 72.5% of patients had satisfactory level of knowledge regarding thalassemia, while 27.5% of patients had unsatisfactory level of knowledge.

Table (4): Illustrated that the mean of physical, psychological, social and environmental domain were (43.75±15.9), (49.9±15.2), (46.83±11.6) and (49.45±11.8) respectively and the mean of overall quality of life and satisfaction with health were (2.4±0.98). While the total mean score of the quality of life domain was (49.33±11.3).

Table (5): Showed that 22.5% of patients had good level of quality of life, while 77.5% of patients had poor level of quality of life.

Table (6): Illustrated that there was a statistical significance difference between total level of knowledge and educational level of patients under study where the lower score for patients who can't read and write and the higher score was for those with primary & secondary education (p= 0.004). In addition, there was a statistical significance difference between total level of knowledge and residence of patients under study where the lower score for patients who live in rural areas and the higher score was for those who live in urban areas (p=0.002). On the other hands there was no statistical significant relation between total level of knowledge and age, gender, marital status, occupation, and income of patients under study where p-value >0.05.

Table (7): Illustrated that there was a statistical significance difference between total quality of life of patients under study and gender where male patients got higher mean score rather than female with p-value= 0.01. In addition, there was a statistical significance difference between total level of quality of life and residence of patients under study where those who live in urban areas got higher mean score rather than who live in rural areas with p-value= 0.04. This table also showed that there was a statistical significance difference between total level of quality of life and income of patients under study where patients with enough income got higher mean score rather than those with low income with p-value= 0.03, while there was no relation between total quality of life scores and age, marital status, educational level, and occupation of patients under study where p-value >0.05.

Table (8): Illustrated that there was no statistical significance relation between total knowledge level and quality of life of patients with thalassemia where p-value 0.6.

Table (1): Frequency and percentage distribution of demographic characteristics of patients under study (n=80).

Patients' Characteristics	Number (N=80)	Percentage (%)
Age		
18-<24	42	52.5%
25-<34	18	22.5%
35-≤45	20	25%
Mean ±SD 23.8 ±6.4		
Gender		
Male	50	62.5%
Female	30	37.5%
Marital status		
Single	58	72.5%
Married	18	22.5%
Education level		
Can't read & write	20	25%
Primary education	36	45%
Secondary education	24	30%
Occupation:		
Work	22	27.5%
Does not work	58	72.5%
If work		
Part time	20	25%
Full time	2	2.5%
Residence		
Urban	22	27.5%
Rural	58	72.5%
Living condition		
living alone	0	0%
living with the family	80	100%
Income		
Enough	22	27.5%
Not enough	58	72.5%

Table (2): Description of patients' total knowledge scores (n=80).

Items	No. of questions	Mean	SD	Range
Blood transfusion therapy	6	4.6	1.1	2-6
Healthy diet	6	4.1	1.5	0-6
Diagnosis of thalassemia	5	3.9	0.72	2-5
Physical activity	4	3.8	0.52	2-4
Complications of thalassemia	5	3.4	1.3	0-5
Iron chelating therapy	6	3.2	2.1	0-6
Treatment of thalassemia	4	3.1	0.84	1-4
Signs and symptoms	5	2.7	1.4	0-5
Definition, cause and types	4	1.7	1.2	0-4
Total knowledge score	45	30.4	7.5	12-44

Table (3): Frequency and percentage distribution of total Knowledge level for patients with thalassemia (n=80).

Knowledge level	No	%
Satisfactory	58	72.5%
Unsatisfactory	22	27.5%

Table (4): Mean & standard deviation of quality of life domains among patients under study (n=80).

Domain	No. questions	Mean	SD	Range
Overall quality of life and general health	2	2.4	0.98	1-10
Physical	7	43.75	15.9	14-82
Psychological	6	49.90	15.2	21-88
Social	3	46.83	11.6	27-67
Environmental	8	49.45	11.8	19-75
Total quality score	26	49.33	11.3	31-77

Table (5): Frequency and percentage distribution of total quality of life among patients under study (n=80).

quality of life level	No	%
Poor	62	77.5%
Good	18	22.5%

Table (6): Relation between total knowledge of the studied patients and their demographic characteristics (n=80).

Patients' characteristics	Knowledge score Mean	SD	F test T test	p-value
Age				
18-<24	30.3	6.6	0.32	0.8
25-<34	29.3	7.8		
35-≤45	31.3	9.2		
Gender				
Male	31.04	6.4	1.06	0.3
Female	29.2	9.1		
Marital status				
Single	31	6.6	1.5	0.2
Married	27.8	10		
Education level				
Can't read & write	25.7	9.3	5.8	0.004*
Primary education	32.3	5.2		
Secondary education	31.3	7.4		
Occupation				
Does not work	30.6	6.7	0.45	0.7
Work (part time)	29.7	9.3		
Residence				
Urban	34.7	3.3	3.2	0.002*
Rural	28.9	7.9		
Income				
Enough	31.1	4.3	0.54	0.6
Not enough	30.1	8.4		

Table (7): Relation between total quality of life of the studied patients and their demographic characteristics (n=80).

Patients' characteristics	Total QoL score		F test T test	p-value
	Mean	SD		
Age				
18-<24	51.6	12.7	2.9	0.06
25-<34	49.3	11.5		
35-≤45	44.4	4.8		
Gender				
Male	51.4	12.9	2.2	0.01*
Female	45.8	6.3		
Marital status				
Single	49.6	12.1	0.28	0.8
Married	49.4	9.8		
Education level				
Can't read & write	45.3	8.1	1.7	0.2
Primary education	50.6	12.9		
Occupation				
Does not work	48.1	11.1	-1.6	0.1
Work (part time)	52.6	11.1		
Residence				
Urban	53.8	11.3	2.1	0.04*
Rural	47.9	10.9		
Income				
Enough	53.6	11.4	2.1	0.03*
Not enough	47.7	10.9		

Table (8): Relation between total score of quality of life and total knowledge level among patients with thalassemia (n=80).

Quality of life	Knowledge level				X^2	P- value
	Unsatisfactory		Satisfactory			
	No	%	No	%		
Poor	16	72.7%	46	79.3%	0.39	0.6
Good	6	27.3%	12	20.7%		

Discussion:

Thalassemia is the most common form of inherited anemia worldwide **(Jaafari, Sadidi, Abdolahinia & Shahesmaeili, 2022)**. The chronic nature of the disease and complications associated with clinical signs of the disease and its treatment make multiple physical, psychological and social problems and effects on the quality of life in these patients. Therefore, assessing knowledge and QoL of these patients leads to better understanding of their specific needs and using more effective care **(Bakhshi, Bahreini, Mirzaei & Kiani, 2018)**.

Socio-demographic characteristics of the patients under the study showed that more than half of patients under study, their age group were between (18-24) years with mean age 23.8 ± 6.4. The short life span is associated with iron overload, which may eventually affect the organs and lead to their failure. These results is similar to **Khodashenas et al., (2021)** who reported that the mean age of the studied patients was 25.78 ± 9.18 in a study titled "Quality of life and related paraclinical factors in Iranian patients with transfusion-dependent thalassemia".

As regards to the gender, the results showed that nearly two thirds of patients were males and more than one third of them were females. These results comes in agreement with **Taheri, Nooryan, Karimi & Zoladl, (2020)** who revealed that more than half of the study subjects were males and more than one third of subjects were females in a study titled "Effect of individual psychotherapy with a focus on self-efficacy on quality of life in patients with thalassemia major".

Regarding to marital status, the study results showed that approximately three quarters of patients were single and less than one quarter of patients were married. These results may be due to that half of patients are young and their age ranged from 18-24, which is not the average age of marriage in Egypt. In addition, that most patients are fear of passing the disease to their children by heredity. These results is in the same line with **Khodashenas et al., (2021)** who revealed that more than three quarters of the participants were single and less than one quarter were married.

Concerning the educational level of the studied subjects, the results of this study illustrated that about one quarter of the studied patients can't read and write, less than half of patients had primary education. This result may be due to that the thalassemia is a chronic disease which affect the patient's physical, psychological and social status of patients and cause severe level of fatigue and pain which interfere with the patient's ability to study and go through their education These results is in accordance with **Dehkordi et al., (2020)** who revealed that less than one quarter of patients was illiterate and more than one quarter had high school education in their study that titled "Effects of aquatic exercise on dimensions of quality of life and blood indicators in patients with beta-thalassemia Major".

Regarding to patients' knowledge about blood transfusion therapy of thalassemia, the present study showed that all patients know that the level of hemoglobin that requires blood transfusion is 7g/dl, in addition to that more than two thirds of them know that dizziness or itching are signs and symptoms that should be reported to the health care team as a result of complications of blood transfusion and more than half of patients know that iron overload is the early complication of blood transfusion. This result might be due to frequent blood transfusion that patients had from the beginning of the disease.

This result is supported by **Kalra, Kaur, Sodhi & Kaur, (2019)** who revealed that all patients know that reactions to the blood being transfused and that iron overload could be a potential complications of repeated transfusions in a study that titled "Knowledge, attitude and practice of transfused thalassemia patients regarding thalassemia in thalassemia day care unit in government medical college, Amritsar, Punjab, India".

In relation to patients' knowledge about diet of thalassemia, the present study demonstrated that the most of patients know that the foods rich in iron like red meat and liver must be avoided for patients with thalassemia. In addition the majority of them know that drinking a cup of tea will inhibit iron the absorption when taking a meal containing iron. This result may be due to increase patients' awareness by the medical staff about the proper nutrition of the thalassemia patient to avoid excessive iron accumulation.

This result go in the same line with **Man, et al., (2019)** who reported that most of the respondents are apart from having the knowledge of foods high in iron content such as liver, beef, oyster and clam, and the inhibitory effects of tea on iron absorption in a study that titled "Knowledge, attitude and practice towards dietary iron among patients with thalassemia and their caregivers in Peninsular Malaysia".

Concerning patients' knowledge regarding activity of thalassemia, the present study illustrated that all patients know that the walking was a type of physical activity that is appropriate for patients with thalassemia and dyspnea and joint pain are symptoms that increased with physical activity. In addition most of them know that decrease hemoglobin level will affect the level of physical activity done by patients with thalassemia. This

result may be due to that thalassemia patients suffering from anemia and low hemoglobin level is one of its main signs which affects the level of physical activity for those patients.

The present result disagree with **Kharyal et al., (2021)** who revealed that less than half of patients know that physical activities of thalassemia patients are affected by the level of hemoglobin and dyspnea aggravated by daily activity in a study that titled "Disease knowledge and general self-efficacy among adolescents with thalassemia major and their parents' perspective".

Regarding the total score of patients' knowledge in the study, this result revealed that nearly three quarters of patients had a satisfactory level of knowledge regarding thalassemia. This result might be due to that thalassemia is a chronic disease, which begins in the childhood and requires the patients to visits the hospital frequently for treatment and follow-up, which makes the patient more experienced with the nature of the disease. In the same context **Asa et al., (2021)** found that the knowledge of the respondents about thalassemia was good in a study that titled "Empowering thalassemia patients and family to increase public knowledge on thalassemia".

As regards to physical domain of quality of life, the result of the current study revealed that nearly two thirds of patients had a little level of pain and discomfort, more than one third of patients had satisfied level of quality of sleep and rest and less than half of patients neither satisfied nor dissatisfied about activities of daily living. This result may be due to the effect of chronic disease over time because the disease started in the childhood period for patients under study, which negatively affects the patients' physical abilities.

This result is similar to **Miladinia, Baraz, Ramezani & Malehi, (2018)** who reported that majority of patients with thalassemia had poor quality of life and suffered from pain, fatigue and sleep disorders in study that titled "The relationship between pain, fatigue, sleep disorders and quality of life in adult patients with thalassemia". Also the result agree with **Salehipour, Ghaljeh, Navidian & Sarani., (2021)** who demonstrated that patients had impaired in physical domain in all items in a study that titled **"**Impact of continuous care model on the quality of life of patients with thalassemia major**"**.

In relation to psychological domain of quality of life, the result of the current study revealed that half of patients had quite often negative feelings such as anxiety and depression toward their quality of life and less than half of them had that a little level of satisfaction regarding bodily image, appearance and self-esteem. This result may be due to the long term effect of the disease that cause their feeling of disabilities and fear from the future.

The result is disagree with **Rikos, Giannadaki, Spontidaki, Tzagkaraki & Linardakis, (2021)** who reported that one quarter of patients had some form of anxiety and few of them presented with some type of depression in a study that titled "Health status, anxiety, depression, and quality of life of patients with thalassemia". While the result was agree with **Shawkat eet al., (2019)** who reported that the thalassemic facies, poor body image, negative feelings and poor self-esteem in adult thalassemia patients had a negative impact on psychological domains in study that titled "Evaluating health-related quality of life (HRQoL) in Iraqi adult patients with beta-thalassemia major using two different iron chelation therapies".

Concerning social domain of quality of life, the result of the current study revealed that more than half of patients had dissatisfied level of social support and less than half of patients had satisfied regarding personal relationships. This result may be due to the effect of the disease on patients that requires frequent hospitalization due to blood transfusion and iron chelation therapy, which limited the interaction of patients with other community members. Also phenotypic features and physical difficulties caused by complications

of thalassemia can make them to withdraw from their social lives.

The result is inconsistent with **Floris et al., (2018)** who reported that the higher scores represented in the social domain may suggest good social integration and the presence of appropriate social support in a study that titled "Quality of life in Sardinian patients with transfusion-dependent thalassemia".

Regarding to the environmental domain, more than half of patients had a moderate level of quality of life regarding physical environment (pollution / noise / traffic / climate) and approximately half of patients' dissatisfied level of satisfaction regarding health and social care accessibility and quality, home environment and financial resources. This result may be due to that most of the patients under study were from the rural area which is far from the central hospitals, and the patients have difficulty in the transport to go to hospital for follow-up and treatment. Also, the insufficient income of patients under study causes inadequacy of housing for living and feeling of insecurity due to the inability of patients to meet their basic needs.

This finding is supported by **Bakhshi et al., (2018)** who reported that the environmental domain that assess the physical environment, financial resources, freedom, physical safety, and home environment got low scores in a study that titled **"**The effect of group counseling on the quality of life in patients with major thalassemia referred to the thalassemia treatment center in Bushehr".

Regarding total score of quality of life for patients under study, this study revealed that more than three quarters of patients had poor quality of life. This might be due to early-onset of the disease in the childhood period and over time, the quality of life of patients was affected due to the absence of self-care concepts, among patients under study.

This finding is supported by **Nashwan et al., (2018)** who reported that patients with thalassemia major got lower score in overall QoL in a study that titled "Quality of life among adolescents aged 14 to 18 years with beta-thalassemia major". While, the finding of this study is inconsistent with **Floris et al., (2018)** who reported that the adults with transfusion-dependent thalassemia obtained scores suggestive of a good quality of life in all areas investigated with the WHOQoL-BREF questionnaire.

Regarding the relation between demographic characteristics and knowledge among patients under study, this study revealed that there was a statistical significance relation between total level of knowledge of patients under study and their educational level and residence. This study might be due to the high level of education in urban areas, which promote education of patients to learn more about the disease.

This result is supported by **Kharyal et al., (2021)** who revealed that the knowledge of the adolescents and their parents showed significant association with the educational level and residence, adolescents and their parents, who were educated to secondary level or above and residing in urban area had better knowledge as compared to those educated till middle level and residing in rural area.

In addition, there was no statistical significant relation between total level of knowledge of patients under study and their age, gender, marital status, occupation and income. This result is in the same line with **Asa et al., (2021)** who reported that the sociodemographic factors such as age, gender, residence, and economic factors were not associated with knowledge of patients with thalassemia in a study that titled "Empowering thalassemia patients and family to increase public knowledge on thalassemia".

Regarding the relation between demographic characteristics and quality of life of patients under study, this study revealed that there was statistical significance relation between quality of life of patients under study and their gender, residence and income. This might be due to that thalassemia treatment hospitals are very few

in number and where located only in capital of Egypt. Patients face many difficulties to travel from their cities to capital many times for follow up and treatment as blood transfusions.

On the other hand, those patients who are living in urban areas have access to transfusion centers and face low difficulties. So their quality of life was found to be better. Regarding to gender difference, it may be due to that the female have more responsibilities and duties to do at home based on their roles as a mother which affect their health and low of time to care of themselves. So, they had poor quality of life than male patients.

This result is disagree with **Khan, Nawaz, Asif & Rehman, (2019)** who reported that there is significant association with residential address of patients. Patients living in rural areas had low quality of life scores in a study that titled "Quality of life of thalassemia major patients". While, the result was agree with **Patel et al., (2019)** who reported that female gender was associated with lower quality of life than male gender. Several reasons have been proposed for this gender difference, including genetic predisposition, increase responsibility and vulnerability to stress in a study that titled "Depression and quality of life in patients with thalassemia in SriLanka".

On the other hands, there was no statistical significant relation between quality of life and age, marital status, educational level, and occupation of patients under study. This might be due to the fact that the adult patients are aware enough to adjust and cope with their disease and the disease causes a physical, psychological and social burden, regardless of marital status or educational level.

The result is similar to **Khodashenas et al., (2021)** who reported that there is no significant correlation between QoL domains and age and marital status, which may be due to the small sample size and other demographic features such as age and marital status are virtually similar among the study population., while the result is disagree with **Khodashenas et al., (2021)** who reported that the patients who having higher education was correlated with higher psychological and social QoL scores.

Regarding relation between quality of life and total knowledge level among patients with thalassemia, the study illustrated that there was no statistical significance relation between total knowledge level and quality of patients under study. This result might be due to that thalassemia is a chronic disease, which started in the childhood period make the patients over time more experienced with the nature of the disease but due to constant fatigue and frequent hospitalization for treatment and follow-up are adversely affected the patients' physical, psychological and social abilities. So, the patients had poor quality of life.

Thalassemia is the most common hereditary disease in the world and in Egypt. The chronic nature of the disease and complications associated with clinical signs of the disease and its treatment make multiple physical, psychological and social problems and effects on the quality of life in these patients.

Conclusion:

Based on the findings of this study it can be concluded that:

- More than two thirds of patients had satisfactory level of knowledge regarding thalassemia.

- The study showed that more than three quarters of patients had poor quality of life.

- The current study illustrated that there was no statistical significance relation between total knowledge level and level of quality of life for patients with thalassemia.

Recommendation

Based on the findings of the present study, the following are recommendations are suggested:

Recommendations related to patients:

- Regular follow up for all patients with thalassemia to evaluate their health conditions and to detect complications early.

- Simple booklet written in an Arabic language should be developed, for patients with thalassemia and includes all needed information

Recommendations related to researchers:

- Replication of the study on a large probability sample selected from different geographical areas in Egypt is recommended to obtain more generalized data.
- Further research studies are needed to focus on studying factors affecting quality of life of patient with thalassemia.

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