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Effect of Self -Determination Theory Based Intervention on Quality of Life Among School Age Children with Thalassemia

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Abstract: Background: Self- determination theory (SDT) play an integral role in the management of many children with thalassemia. **Purpose:** The purpose of the study was to assess the effect of the self- determination theory based intervention on quality of life among school age children with thalassemia. **Research Design:** A quasi experimental design was used. **Setting:** It was conducted the pediatric thalassemia unit in Menoufia University Hospital. **Sample:** Purposive sample of 70 children with thalassemia at the previously mentioned setting. **Instruments:** Three data collection instruments were used: structured interviewing questionnaire, Basic Psychological Needs Satisfaction and Frustration Scale and Standardized QOL checklist. **Results:** the result of this study showed that there were very highly statistical significant differences between children's knowledge on posttest compared to pretest (3.6±1.1 Vs 1.7±0.6 respectively). Also, children who received self- determination theory based intervention had improved their autonomy, relatedness, competency and quality of life dimensions. **Conclusion:** It was concluded that implementation of self- determination theory improved knowledge and quality of life dimensions for children with thalassemia on post and follow-up tests than pretest. **Recommendation:** self- determination theory based intervention about thalassemia should be developed and provided for children in pediatric thalassemia units.

Key words: *Self- Determination Theory, Self-care, Thalassemia.*

Introduction

Thalassemia is the most common hemoglobin disorder in the world. It is an inherited disease, where if the parents are carriers or affected, their children will have high probability of being affected too.

It is one of health problem, worldwide it occurs in 4.4/10,000 live births. About 66.0% of children were under 15 years old. It affects approximately 200 million children worldwide. Globally, there are about 240 million carriers of β-thalassemia. In the United States, it was reported that 1: 272, 000 live births had thalassemia (Sahmoud et al., 2020).

In Egypt, β-thalassemia is considered the most common monogenic disorder with a carrier rate of almost 5.3 to 9.0 %, representing the most common genetically determined chronic hemolytic anemia (85.1%) and 1–5 million neonates are anticipated to be affected with this illness, (Ragb et al., 2021). A high rate of carriers has been

reported ranging from 4–5% reaching up to 9–10% (Shanshory et al., 2021).

Thalassemia has an effect on the formation of the globin chain. Signs symptoms may vary depending on the number and type of chains involved (Safizadeh et al., 2012). Thalassemia usually accompanied by physiological changes in the body structures of children with severe diseases related to hemochromatosis such as growth retardation and delayed or absent sexual maturation in many adolescents. Moreover, most children treated with blood transfusion and early chelation therapy survives well into adulthood. The most common causes of death are heart diseases, post- splenectomy sepsis, and multi organ failure secondary to hemochromatosis (Wilson et al., 2011).

Management strategies of beta-thalassemia major emphasis on just how to treat its manifestations and chronic complications using the best current treatment approaches, transfusion therapy, oral iron chelation therapy, splenectomy, , in addition to hematopoietic stem cell transplantation (Eliezer et al., 2016. Transfusion requirements increase with time and children develop iron overload for which children require chelation (Naggarwal et al., 2011). Blood transfusion aims to promote normal growth and chelating agents are capable of removing excessive iron from the body (Navaneetha et al., 2013).Thalassemia has no cure, so the goal of treatment is to normalize the hemoglobin and hematocrit of the child, thus alleviating the symptoms of severe anemia. This is accomplished via a regular schedule of transfusions, with many children requiring transfusions every 2 to 3 weeks (Potts et al., 2012).

Health related quality of life is an important part for children health. It determines the ability to manage symptoms, physical, psychological outcomes, lifestyle changes, behavior of taking medications as well as following diets and instructions (Eiser &Varni, 2013).

Self-determination theory (SDT) is an empirically based theory of motivation and psychological development that is especially focused on the basic psychological needs (autonomy, competence and relatedness) that promote high quality motivation and wellness, and how they are supported in social contexts. Thalassemia children need to regulate their own actions regarding his condition, development of securing and satisfying connections with others, adhere to treatment regimen and maintain a healthy lifestyle to have better self-care (Ryan & Deci (2017).

Significance of the study

Thalassemia is an inherited blood disease. It is a serious health problem throughout the Mediterranean region, the Middle East and the Southeast Asia (Behdani et al., 2015).There is approximately 300 million carriers of this hemoglobin disorder worldwide (Kavitha &Padmaja, 2017). In Egypt, thalassemia is the most common genetically inherited hemoglobin disorder with a carrier rate ranging from 0.5% to more than 9% and 1000 children out of 1.5 million live births are born annually with thalassemia major (De Sanctis et al., 2017).

School age children with thalassemia want to have more control over their body, they ask many questions about procedures, investigations and treatment (Wilcox &Verhovsek, 2012). Limited studies were conducted in this aspect. Therefore, this study will assess the effect of self- determination theory based intervention on self- care and quality of life among school age children with thalassemia.

Purpose

The purpose of this study is to assess the effect of the self- determination theory based intervention on quality of life among school age children with thalassemia.

Research Hypothesis:

- Children with thalassemia who receive the self-determination theory based intervention will have higher level of knowledge on posttest than pretest.
- Children with thalassemia who receive the self-determination theory based intervention will have higher level of autonomy, relatedness and competency on posttest than pretest.
- Children with thalassemia who receive the self-determination theory based intervention will have higher quality of life on posttest than pretest.

Methods

Research Design:

A quasi- experimental design (pre and posttest) was utilized for this study.

Sampling:

Purposive sample of 70 children with thalassemia at previously mentioned setting and who agree to participate in the study and meet the criteria of sample selection will be included).

Instruments:-

Two instruments were utilized for data collection:

Instrument one: A structured interviewing questionnaire to assess general characteristics of the studied sample:

It was developed by the researcher guided by Zaghamir et al., (2019). It consisted of two parts:

- **Part one:** Socio-demographic characteristics of studied children. It included questions about age, gender, diagnosis, Suffer from other diseases, Consanguinity with parents, family history of thalassemia, family history of blood diseases, time of starting treatment, hemoglobin level, number of blood transfusion / year and spleen removal.
- **Part two:** Children knowledge about thalassemia. It included knowledge assessment items such as: definition of thalassemia, causes, types, symptoms, Management and prevention of thalassemia.

Total Scoring system (0-5):

Scoring items	*Score*
Good knowledge (4-5)	> 75%
Fair knowledge (3)	≤75%
Poor knowledge (0-2)	≤ 50%

Reliability of Instrument 1:

Reliability was estimated by using test retest method with two weeks apart between them. Then Cronbach alpha was calculated between the two scores using SPSS computer package. It was 0.85 which indicates that the instrument is reliable for the study.

Instrument 2: Basic Psychological Needs Satisfaction and Frustration Scale (BPNSFS):

It was adopted from Ryan & Deci (2000) & Vansteenkiste, et al., (2015) to assess child's autonomy, relatedness and competency either satisfaction or frustration. Each item contains 4

questions and rated on a 4-point Likert scale.

Total scoring system for each item (4-16):

Scoring items	*Score*
High autonomy satisfaction (13 – 16)	≥ 60%
Moderate autonomy satisfaction (10-12)	30% to < 60%
Low autonomy satisfaction (4-<10)	< 30%

Reliability of Instrument 2:

Reliability was estimated by using test retest method with two weeks apart between them. Then Cronbach alpha was calculated between the two scores. It was 0.81 which indicates that the instrument is reliable for the study.

Instrument 3: Standardized QOL checklist:

It was adopted from Ware & Sherburne (1992). It consisted of six dimensions with 29 items such as general health (6 items), Limitation of daily activity (9 items), Physical health problems (4 items), Emotional health problems (3 items), pain perception (1 item), Energy and emotions (6 items). Each item is rated on a 3-point Likert scale. Domain scores were scaled in a positive direction (i.e., higher scores donate higher QOL)

Total scoring system (29-87)

Scoring items	*Score*
Good QoL (66-87)	≥ 80%
Average QoL (50-65)	60% to <80%
Poor QoL (29-49)	< 60%

Reliability of Instrument 3

Reliability was estimated by using test retest method with two weeks apart between them. Then Cronbach alpha was calculated between the two scores using SPSS computer package. It was 0.87 which indicates that the instrument is reliable for the study.

Validity

For validity assurance, instruments were submitted to a jury of five experts in the Medical and Nursing field (3 professors in nursing and 2 professors in medicine) to modify any required items of the instruments .The modifications were done to ascertain their relevance and completeness.

Ethical considerations

- An initial approval was obtained from the Ethical Research Committee in the Faculty of Nursing, Menoufia University
- A written consent was obtained from the parents of children who will participate in the study.
- An initial interview done to inform children and their parents about the purpose, benefits of the study and explain that participation in the study was voluntary and the participants can withdraw from the study at any time without penalty.

Pilot study

It was carried out on 7 children (10% of the sample) after the instruments were developed and before starting the data collection to test the practicability, applicability and to estimate the needed time to fill the instruments.

Procedure

Assessment phase:

1) Prior to data collection, a written permission to carry out the study was obtained from the director of the unit after submitting an official letter from the Dean of the Faculty of Nursing at Menoufia University explaining the purpose of the study and methods of data collection.

2) Data collection for this study was conducted over a period of five months extending from 1st of September, 2021 to the end of February, 2022.

3) The researcher introduced herself to the studied children and their parents. The purpose of the study and methods of data collection was explained to them.

4) The researcher interviewed each child and fulfilled the structured interviewing questionnaire regarding their knowledge about thalassemia using instrument 1. It took 15 minutes to fulfill (pretest).

5) Instrument 2 was used by the researcher to assess children basic psychological needs (autonomy, relatedness and competency) (pretest).

6) Assessment of children health related quality of life was assessed by the researcher using instrument 3(pretest)

Implementation phase:

- Based on assessment of children knowledge weakness and practice defects the intervention based on Self-determination theory was designed accordingly.
- Health teaching sessions were started by the researcher to all children with thalassemia at the Pediatric Thalassemia Unit. Each session included 7-8 children. Each session lasted for 20 to 30 minutes. Oral presentations, group discussions, smart phone, demonstration and re-demonstration and feedbacks were used for health education also, explanatory booklets were distributed between children.
- The first session contained theoretical knowledge related to thalassemia e.g. definition, causes, types, signs, symptoms and complications of thalassemia. It lasts about 30 minutes.
- The second session contained theoretical knowledge related to management of thalassemia, benefits and complications of blood transfusion. It lasts about 30 minutes.

➢ The researcher provided summary about knowledge provided in the first session. Afterwards, nursing care interventions based on Self-determination theory about improving quality of life were discussed.

➢ Direct reinforcement was used in the form of chocolates and pens were distributed between children.

➢ Also each session ended by a summary of its contents and feedback from the children was obtained to ensure that they got the maximum benefit.

➢ Explanatory booklet that used was translated into Arabic by the researcher and was distributed to all children.

Evaluation phase:

1) Reassessment of children knowledge regarding thalassemia was done immediately following the health education sessions by the researcher to each child individually within the group using instrument 1 (posttest).

2) Reassessment of children's health related quality of life was done to each child individually within the group after 30 days following the health education sessions by the researcher using instrument 3(posttest).

3) Reassessment of children' knowledge, as well as quality of life were done three months later to each child individually within the group by the researcher using instrument 1 & 3 (follow up test).

Statistical analysis:

Data was entered and analyzed by using SPSS (Statistical Package for Social Science) statistical package version 22. Graphics were done using Excel program.

Quantitative data were presented by mean (X) and standard deviation (SD). It was analyzed using student t- test for comparison between two means, and ANOVA (F) test for comparison between more than two means.

Qualitative data were presented in the form of frequency distribution tables, number and percentage. It was analyzed by chi-square ($\chi 2$) test. However, if an expected value of any cell in the table was less than 5, Fisher Exact test was used(if the table was 4 cells) , or Likelihood Ratio (LR) test (if the table was more than 4 cells). Level of significance was set as P value <0.05 for all significant tests.

Results

Table 1:- showed characteristics of studied children. It was obvious from this table that 35.7% of the studied children with thalassemia aged between 6 to 8 years with mean age 9.1 ± 1.8 years and more than half were females (55.7%).

Table 2:- reveals Medical data of studied children. As illustrated in the table, nearly two third of children diagnosed with thalassemia since more than 5 years (64.3%), with a mean duration of 6.2 ± 1.5 years, and nearly half of them had Consanguinity with parents (45.7%). Also, approximately one third had family history of thalassemia (28.6%).Meanwhile; more than half of children had low hemoglobin level before blood transfusion (58.6%) and the majority of them had spleen removal.

Table3:- displays Children knowledge about thalassemia on pre, post, and follow up tests. The findings revealed that children had higher level of knowledge about thalassemia on post and follow-up tests than on pretest. Therefore, there were highly statistical significant differences between children 'knowledge on posttest than pretest regarding definition, causes, types, symptoms, management and prevention of thalassemia (P<0.0001).However, there were no statistical significant differences regarding children 'knowledge between posttest and follow-up test.

Table 4:- displays Mean total score of children knowledge on pre, post and follow up tests. The findings revealed that there were highly statistical significant differences between pretest and posttest (P<0.0001).Also, there were highly statistical significant differences between pretest, posttest and follow- up test (P<0.0001). However, there were no statistical significant differences between posttest and follow-up test (P >0.05).

Table 5:- displays Assessment of thalassemic children's Autonomy level of satisfaction and frustration on pre, post and follow-up tests based on self-determination theory. The findings revealed that there were highly statistical significant differences regarding levels of autonomy satisfaction on pre, post and follow up tests (P<0.001).Also, there were highly statistical significant differences regarding levels of autonomy frustration on pre, post and follow up tests (P<0.001).

Table 6:- displays Assessment of thalassemic children's Relatedness level of satisfaction and frustration on pre, post and follow up tests based on self-determination theory. The findings revealed that there were statistical significant differences regarding levels of relatedness satisfaction on pre, post and follow up tests (P<0.05)

Table 7:- displays Assessment of thalassemia children's Competency on pre, post and follow up tests based on

self-determination theory. The findings revealed that there were statistical significant differences regarding levels of Competency satisfaction on pre, post and follow up tests

Table 8 :- displays total scores of quality of life dimensions on pre, post and follow up tests. As shown in this table the findings revealed that there were highly statistical significant differences in total physical health problems, total energy and emotional health and grand total QoL on pre, post and follow-up tests (P<0.001). Also, there were statistical significant differences in total emotional health problems (P <0.05) on pre, post and follow-up tests.

Table 9 and Fig.1:- highlight Total quality of life levels on pre, post and Follow tests. Posttest program revealed a highly significant improvement (p<0.0001) in the quality of life. Post program' good quality of life increased from 21.4 % on pretest to 40% on posttest and the difference was highly significant (P<0.0001). In addition, poor quality of life was decreased from 61.4% on pretest to 17.1% on posttest, and to 15.7% on follow- up test. Therefore there were highly statistical significant in post program intervention (P<0.0001). Moreover, the mean score of total quality of life increased from 43.8 ± 11.7 on pretest to 50.1 ± 9.3 on posttest, and 48.2 ± 9.6 on follow -up test. So, the difference was highly significant (P<0.0001)

Table (1): Characteristics of studied children (N = 70).

Children characteristics	N0 70	%
Age (Years)		
6 – 8 years	25	35.7
9 – 10 years	23	32.9
11 – 12 years	22	31.4
Mean ± SD= 9.1 ± 1.8 years (Range=6-12 years)		
Gender		
Male	31	44.3
Female	39	55.7
Total	**70**	**100**

Table (2): Medical data of studied children (N = 70).

Medical data	N0 70	%
Duration since diagnosed with thalassemia:		
1- 3 years	7	10
4 -6 years	18	25.7
> 6 years	45	64.3
Mean ± SD	**6.5 ± 2.1 years**	
Suffer from other diseases:	No	%
No	100	10
Yes	0	0
Consanguinity with parents:	32	45.7
Yes	38	54.3
No		
Family history of thalassemia:	20	28.6
Yes	50	71.4
No		
Time of starting treatment:	28	40
≤ 5 years	42	60
> 5 years		
Child hemoglobin level before blood transfusion:	41	58.6
< 7 mg/dl	20	28.5
7 – >8 mg/dl	9	12.9
8- 9 mg/dl		
Number of blood transfusion / year:	6	8.6
Every 15 days	41	58.5
Every 30 days	23	32.9
Every 45 days		
Spleen Removal	45	64.3
Yes	25	35.7
No		
Total	**70**	**100**

Table (3): Children knowledge about thalassemia on pre, post, and follow up tests (N = 70).

Children's knowledge about thalassemia	Pretest		Posttest (Immediate-posttest)		P1	Follow-up Test		P2
	No 70	%	No 70	%		No 70	%	
Definition of thalassemia								
Incorrect answer	25	35.7	6	8.6	$\chi2$ =14.9	8	8.6	$\chi2$ =1.1
Correct answer	45	64.3	64	91.4	P < 0.0001 HS	62	91.40	P = 0.23 NS
Causes of thalassemia:								
Incorrect answer	47	67.1	28	40	$\chi2$ =10.4	31	44.3	$\chi2$ =0.12
Correct answer	23	32.9	42	60	P < 0.0001 HS	39	55.7	P = 0.73 NS
Types of thalassemia:								
Incorrect answer	56	80	30	42.9	$\chi2$ =20.4	36	51.4	$\chi2$ = 0.71
Correct answer	14	20	40	57.1	P < 0.0001 HS	34	48.6	P= 0.39 NS
Symptoms of thalassemia:								
Incorrect answer	33	47.1	30	42.9	$\chi2$ =3.6	32	45.7	$\chi2$ = 2.3
Correct answer	37	52.9	40	57.1	P=0.7 NS	38	54.3	P = 0.9 NS
Management of thalassemia:								
Incorrect answer	47	67.1	20	28.6	$\chi2$ =20.9	25	35.7	$\chi2$ =2.5
Correct answer	23	32.9	50	71.4	P < 0.0001 HS	45	64.3	P = 0.11 NS
Prevention of thalassemia:								
Incorrect answer	52	74.3	17	24.3	$\chi2$ =35.1	24	34.3	$\chi2$ =1.2
Correct answer	18	25.7	53	75.7	P < 0.0001 HS	46	65.7	P= 0.26 NS
Services offered to you								
	15	21.4	10	14.2	$\chi2$ =0.6	12	17.1	$\chi2$ =0.10
No								P = 0.70 NS
Yes	55	78.6	60	85.7	P = 0.43 NS	58	82.9	
Kind of services (N=55):	16	22.9	14	20		14	20	
Fund	14	20	17	24.3	$\chi2$ =0.4	17	24.3	$\chi2$ =0.5
Health	7	10	4	5.7	P = 0.42 NS	6	8.5	P=0.43 NS
Social	33	47.1	35	50		33	47.1	
All								
Total	70	100	70	100		70	100	

Table (4): Mean total score of children knowledge on pre, post and follow up tests.

Mean total score	Pretest	Posttest (Immediate-posttest)	P1	Follow-up test	P2	P3
Mean total score of children knowledge	-1.7±0.6	3.6±1.1	t =12.6 p< 0.0001 HS	3.3±1.1	t=1.2 p2=0.14 NS	F=43.5 P3<0.0001 HS

Table (5): Assessment of thalassemic children's Autonomy level of satisfaction and frustration on pre, post and follow-up tests based on self-determination theory.

Basic Psychological Needs: (Autonomy)	Intervention groups						Test of significance
	Pretest		Posttest (after 30 days)		Follow -up test		
	No (70) %		No (70) %		No (70) %		
Groups of Total Autonomy Satisfaction							
Low Autonomy Satisfaction (<10)	52	74.3	35	50	46	65.7	LR=9.6
Moderate Autonomy Satisfaction (10 - 12)	7	10	22	31.4	11	15.7	P<0.002
High Autonomy Satisfaction (13 - 16)	11	15.7	13	18.6	13	18.6	HS.
Groups of Total Autonomy Frustration							
Low Autonomy Frustration (<10)	40	57.1	45	64.3	42	60	LR=14.3
Moderate Autonomy Frustration (10 - 12)	20	28.6	10	14.3	13	18.6	P<0.007
High Autonomy Frustration (13 - 16)	15	21.4	15	21.4	15	21.4	HS
Total	**70**	**100**	**70**	**100**	**70**	**100**	

Table (6): Assessment of thalassemic children's Relatedness level of satisfaction and frustration on pre, post and follow-up tests based on self-determination theory.

Basic Psychological Needs: (Relatedness)	Intervention groups						Test of significance
	Pretest		Posttest (after 3 days)		Follow up- test		
	No (70) %		No (70) %		No (70) %		
Groups of Total Relatedness Satisfaction							
Low Relatedness Satisfaction (<10)	49	70	31	44.3	42	60	χ^2 =3.3
Moderate Relatedness Satisfaction (10 - 12)	17	24.3	33	47.1	22	31.4	P=0.04
High Relatedness Satisfaction (13 - 16)	4	5.7	6	8.6	6	8.6	S
Groups of Total Relatedness Frustration							
Low Relatedness Frustration (<10)	40	57.1	47	67.1	40	57.1	χ^2 =0.7
Moderate Relatedness Frustration (10 - 12)	15	21.4	10	14.3	15	21.4	p=0.40
High Relatedness Frustration (13 - 16)	15	21.4	13	18.6	15	21.4	NS
Total	**70**	**100**	**70**	**100**	**70**	**100**	

Table (7): Assessment of thalassemiac children's Competency level of satisfaction and frustration on pre, post and follow-up tests based on self-determination theory.

Basic Psychological Needs (Competency)	Intervention groups						Test of significance
	Pretest		Posttest (after 30 days)		Follow up- test		
	No (70) %		No (70) %		No (70) %		
Groups of Total Competence Satisfaction							
Low Competence Satisfaction (<10)	50	71.4	40	57.1	42	60	χ^2=2.3
Moderate Competence Satisfaction (10 - 12)	10	14.3	15	21.4	14	20	P=0.01
High Competence Satisfaction (13 - 16)	10	14.3	15	21.4	14	20	S.
Groups of total Competence Frustration							
Low Competence Frustration (<10)	53	90	57	71.4	55	78.6	LR=11.5
Moderate Competence Frustration (10 -12)	10	5.7	7	25.7	8	11.4	P<0.02
High Competence Frustration (13 - 16)	7	10	6	8.6	7	10	S.
Total	**70**	**100**	**70**	**100**	**70**	**100**	

Table (8): Total scores of quality of life dimensions on pre, post and follow up tests.

Total score	Pretest	Posttest (after 30 days)	Follow- up Test	F test	P value
Total general QoL					
Mean ± SD	2.7± 1.4	2.8 ± 1.3	2.7 ± 1.4	0.32	0.72 NS
Range(minimum -maximum)	1 – 6	1-6	1-6		
Total Limitation of daily activities					
Mean ± SD	19.7 ± 4.1	18.3 ± 3.9	19.2 ± 4	2.3	0.11 NS
Range(minimum –maximum)	9-27	9-27	9 – 27		
Total physical health problems					
Mean ± SD	8.6 ± 2.7	7.2± 3.1	8.3±2.7	4.9	<0.008 HS
Range(minimum –maximum)	4-12	4-12	4-12		
Total emotional health problems					
Mean ± SD	6.6±2.0	5.5±2.4	6.3±2.0	4.7	<0.01 S
Range(minimum –maximum)	3-9	3-9	3-9		
Total energy and emotional health					
Mean ± SD	10.1 ±3.9	12.4 ±3.3 ±	11.8 ±3.1	8.5	<0.0001 HS
Range(minimum –maximum)	6 – 18	6 – 18	6 – 8		
Grand total QoL					
Mean ± SD	43.8 ± 11.8	50.4 ± 9.11	48.2 ± 9.6	7.97	<0.0001 HS
Range(minimum –maximum)	27 – 62	30 -70	30 -70		

Table (9): Total quality of life levels on pre, post and Follow tests (N=70)

Total QoL levels	pretest		Posttest (after 30 days)		Test of Sig.	P1	Follow-up test		Test of Sig.	P 2
	No 70	%	No 70	%			No 70	%		
Poor QoL (≤ 49)	28	40	6	8.6	χ2=16.8	< 0.0001 HS	17	24.2	χ2= 4.2	0.05 NS
Average QoL (50- 65)	27	38.6	36	51.4			30	42.9		
Good QoL (66 – 81)	15	21.4	28	40			23	32.9		
Total	70	100.0	70	100			70	100.0		
X± SD	43.8 ± 11.7		50.1 ± 9.3				48.2± 9.6		F test=6.9	P3 < 0.001 HS
Total	**70**	**100.0**	**70**	**100**			**70**	**100**		

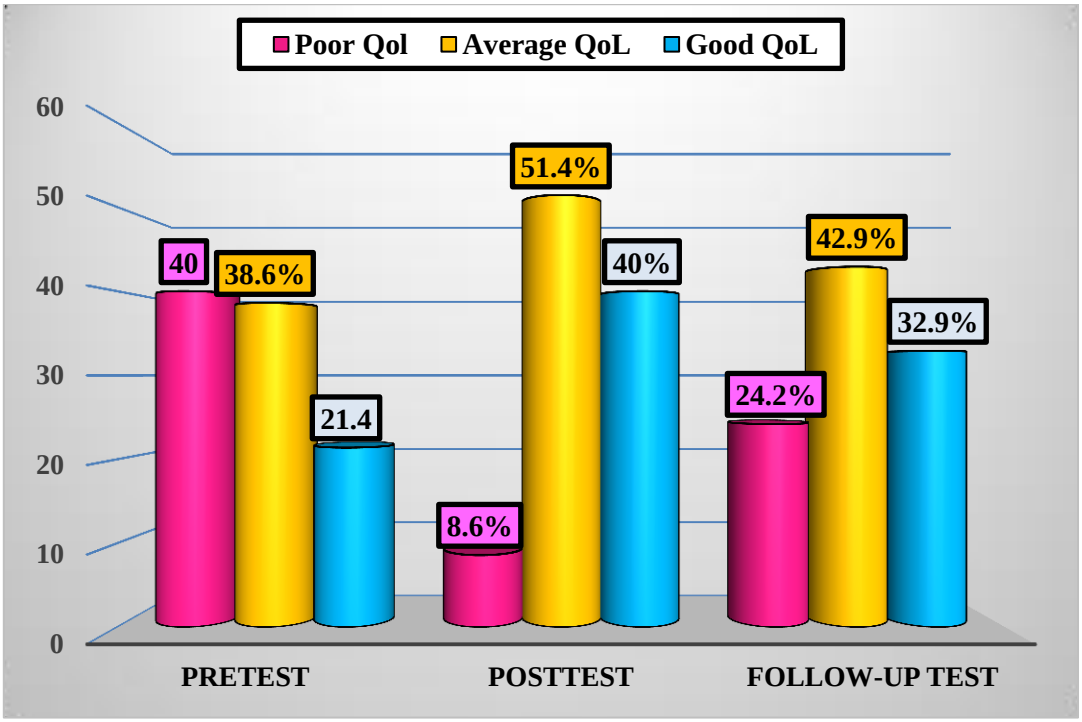

Fig.1: Total quality of life levels on pre, post and Follow tests.

Discussion

Thalassemia is a chronic disease that has a significant impact on the health of children. It necessitates regular medical care and usage of medications on a regular basis (Elsoudy et al., 2022). Thalassemia is an inherited disease, meaning that at least one of the parents must be a carrier. It is a group of hemoglobinopathies in which the normal ratio of alpha to beta globin production is disrupted. This abnormal alpha to beta chain ratio causes the unpaired chains to precipitate, leading to destruction of red blood cell precursors in the bone marrow (ineffective erythropoiesis) and in the circulation (hemolysis). Children with thalassemia have variable degrees of anemia and extra-medullary hematopoiesis, which in turn can cause bone changes, impaired growth, and iron overload (Benz et al., 2022).

Thalassemia and its complications affect self-care and quality of life of those children. Therefore, nursing interventions by nurses as essential careers are important (Madmoli et al., 2019).

Implementation of self- determination theory would significantly improve children's quality of life and Self-care to promote their health and welfare to make their lives better (Ryan & Deci, 2017)

The current study hypothesized that Children with thalassemia who receive the self-determination theory based intervention will have higher level of knowledge on posttest than pretest.Also, Children with thalassemia who receive the self-determination theory based intervention will have higher quality of life dimensions on posttest than pretest.

In relation to hypothesis one: Children with thalassemia who receive the self-determination theory based intervention will have higher level of knowledge on posttest than pretest. The present study illustrated children's' knowledge about thalassemia on pre, post and follow-up tests. The study revealed that the highest level of children's 'knowledge was on posttest (3.6±1.1). From the researcher's perspective, this could be attributed to the positive effect of self-determination theory implementation. Also, children were enthusiastic to learn more about thalassemia.

This result was consistent with Meah 2021 et al., in their study about "Assessment of Awareness Among Parents of Children with Thalassemia Major in Bangladesh" who stated that the awareness and knowledge among parents and children with thalassemia were insufficient. They have inadequate knowledge regarding the disease, safe blood transfusion and treatments of associated complications.

Also, this result came in agreement with Mohamed et al., (2021) in their study about" Effect of Empowerment Program on Self-efficacy among Children with Thalassemia." They found that all children had unsatisfactory knowledge about thalassemia before the program implementation, and significant improvement in knowledge of children detected immediate post and one month after the empowerment program.

In addition, the finding of present study was on the same line with Zaghamir et al., (2019) in their study about "Assessment of Thalassemic Children Knowledge about Thalassemia and Iron Chelation Therapy". They concluded that there is an improvement in the children's knowledge after implementing educational training programs. This can be interpreted that nursing educational program was effective in improving children's knowledge.

Bedsides, this finding was consistent with Biswas et al., (2018) in their study about " Knowledge of the caregivers of thalassemic children regarding thalassemia: A cross-sectional study in a tertiary care health facility of eastern India " who concluded that Children and caregivers' knowledge regarding thalassemia was not at all satisfactory.

In the same line, this result agreed with Atshan,, & Aziz (2022) in their study about " Impact of an educational program on knowledge about chelation therapy & nutrition of children with beta thalassemia major ". They showed that education program was highly effective on improving knowledge of children with beta thalassemia major.

In relation to hypothesis two: Children with thalassemia who receive the self-determination theory based intervention will have higher level of autonomy, relatedness and competency on posttest than pretest.The findings of the current study clarified that there was a significant improvement in the children's autonomy, relatedness and competency. From the researcher's perspective, this could be attributed to the clarity and simplicity of the methods of teaching (oral presentations, group discussion, smart phone, communication board, feedbacks and explanatory booklets) that were used in sessions which in turn helped children to acquire and improve their knowledge, autonomy, relatedness and competency. Besides, the utilized communication techniques (e.g. good listening, feedback and two-way communications) could be effective.

Regarding autonomy, the present study concluded that there was significant improvement in children's autonomy score on posttest than on pretest. This result was in line with Nourbakhsh et al., (2021) in their study about "The Association between Behavioral Problems with Self-Esteem and Self-Concept in Pediatric Patients with Thalassemia". They found that children had poor self-esteem and low autonomy.

In addition, these results came in agreement with Tomaj et al., (2016) who conducted a study about" The Effects of Group Play Therapy on Self-Concept Among 7 to 11 Year-Old Children Suffering From Thalassemia Major. They showed that the mean self-concept and autonomy score was significantly higher at the second point in time compared to the baseline. They also showed that group play therapy improved self-concept and autonomy in those children.

Regarding relatedness, the present study concluded that there was significant improvement in children's relatedness score on posttest than on pretest. These results came in agreement with Stanton et al., (2020) in their study about "Self-determination theory in acute child and adolescent mental health inpatient care. A qualitative exploratory study". They showed that Engaging young people in activities with a focus on relatedness, autonomy and competence had therapeutic benefits. Connection with staff and peer interaction improve children's relatedness and enhance their safety.

Also, This result was consistent with El zaree et al., (2018) in their study about "Adaptive Functioning and Psychosocial Problems in Children with Beta Thalassemia Major ". They found that thalassemic children had a relatively mild affection for adaptive and psychosocial functioning that can be explained by social and medical support they receive, which may increase their competence and psychological wellbeing.

Moreover, this result came in agreement with Keshvari et al.,(2013) in their study about " Relation between Children's Well-Being and Family

Function in Children with Thalassemia Major in Isfahan ". They concluded that the children with thalassemia had low relatedness, and there is a direct relationship between family functioning and emotional well-being of children with thalassemia.

Regarding competency, the present study concluded that there was significant improvement in children's competency score on posttest than on pretest. These results came in agreement with Feng et al., (2022) in their study about "Effect of Self-Determination Theory on Knowledge, Treatment Adherence, and Self-Management of Patients with Maintenance Hemodialysis ".They showed that Self-determination theory management is effective in improving hemodialysis-related treatment compliance, self-management ,competency level and quality of life.

Also, these results came in agreement with Baghersalimi et al., (2021) in their study about "Evaluation of Self-efficacy in Children and Adolescents With Thalassemia Major" they showed that the rate of self-efficacy and competency in children with thalassemia was moderate, and the allocation of a specific ward, easy access to health care staff, and social support for children may seem to justify the moderate to good self-efficacy.

Besides, this finding was in line with Wongnonglaeng et al., (2021) in their study about " The effects of health program based on Bandura's theory using gamification on perceived self-efficacy in health care of school-age children with thalassemia " They showed that the mean score of perceived self-efficacy in health care of school-age children with thalassemia in the experimental group at the end of the intervention and at 1 week post intervention was significantly higher than that before the intervention and in the control group.

This finding was also in conformity with Elsoudy et al., (2022) in their study about "Original Article Compliance of Children with Thalassemia to Their Therapeutic Regimen." They concluded that the majority of children with thalassemia were able to comply with their therapeutic regimen.

Furthermore, this finding was in line with Hamamy, & Al-Allawi (2013) who conducted a study about "Epidemiological profile of common haemoglobinopathies in Arab countries" they showed that school-age children had more increased abilities to complete their health care tasks such as filling prescription or recognize symptoms of illness.

In relation to hypothesis Three: Children with thalassemia who receive the self-determination theory based intervention will have higher quality of life on posttest than pretest.The present study illustrated that children with thalassemia had poor QoL in physical, psychological, social, and school functioning domains on pretest, while they had good QOL in posttest. This could be attributed to the poor knowledge of children before implementation of intervention program which was reflected negatively on their QOL and the effect of chronic disease. This also, could be attributed to the effectiveness of the intervention. This result was consistent with Shackleford et al., (2019) in their study about "Applying the Self-determination Theory to Health-related Quality of Life for Adolescents with Congenital Heart Disease". They concluded that relatedness and competence were significantly associated with higher quality of life in those patients.

Also, this result was consistent with Elalfy et al., (2014) in their study about

"Quality of life of Egyptian b-thalassemia major children and adolescents". They concluded that children with thalassemia had a poor QOL on pretest, and conditions like high hemoglobin level and low iron overload were associated with improved QOL scores.

In addition, this result agreed with Hamdy et al., (2021) in their study about "Assessment of Quality of Life among Beta-Thalassemia Major Patients Attending the Hematology Outpatient Clinics at Cairo University Hospital". They reported that QoL in thalassemia major patients is compromised. So, the necessary interventions that focus on the affected quality of life domain should be done to improve their QOL.

Bedsides, this finding was in line with Mahmoud et al., (2019) in their study about "Quality Of Life of Children with Thalassemia in Alexandria ". They reported that Thalassemia is negatively affecting the children's QOL. Where, the majority of the children and their parents reported low QOL and the minority of them had moderate QOL. Meanwhile, small percent of them (8.5%) had high QOL.

Also, this result agreed with Batoolet al., (2022) in their study about "Factors affecting health-related quality of life (HRQoL) in Pakistani children with thalassemia". They reported that children with thalassemia had poor QOL. They also reported that there is a need for improvement in thalassemia management, and a modification of healthcare services improved HRQoL in those patients.

Meanwhile, this result agreed with Mediani al., (2021) in their study about "Predicting factors impact to quality of life of school age Thalassemic children in Indonesia". They reported that effective transfusion scheduling (frequency of blood transfusion), family and friends support, and providing adequate education were effective in improving QOL of thalassemic children in Indonesia.

In addition, the finding of present study was in the same line with Hakeem et al., (2018) in their study about "Health-related quality of life in pediatric and adolescent patients with transfusion-dependent ß-thalassemia in upper Egypt ". They concluded that younger age had better scores regarding social, emotional, psychological and total scores compared to older ones after the program implementation than before its implementation.

Also, this result agreed with Sargolzaei al., (2020) in their study about " Interventions to improve Quality of Life in Patients with Major Thalassemia: A Systematic Review ". They reported that educational interventions had a positive effect on improving the quality of life of Patients with Major Thalassemia.

Besides, this finding was in line with Abu Samra (2015) in their study about "Impact of educational program regarding chelation therapy on the quality of life for Beta-thalassemia major children "They found that there was a positive effect of the educational program in improving children's knowledge score and their quality of life.

Furthermore, this result came in agreement with El -Kamah,et al., (2014) in their study about " Quality of Life Outcomes Among Egyptian Children with Beta-Thalssemia " They concluded that Children with β-thalassemia had low QoL outcome scores, so community programs and parental education are needed to support children with β-thalassemia in developing countries and improve their quality of Life.

In the same context, the study was consistent with Jeesh et al., (2018) in their study about "The Effects of

Patients' and Care-Givers' Knowledge, Attitude, & Practice (KAP) on Quality of Life Among Thalassemia Major Patients' in Damascus-Syrian Arab Republic ". This study revealed that a remarkable improvement in both knowledge and performance was found after patients received the teaching guide, which lead to an increase in overall patient's quality of life.

In addition, this result agreed with Eldakhakhny (2011) in their study a bout "Quality of Life of School Age Thalassemic Children at Zagazig City ". They reported that thalassemia has a negative impact on perceived physical, emotional, social and school functioning in thalassemia patients. So that programs should be developed to increase children's adherence to the treatment regimen, increase psychosocial support and improve their QOL.

Also, this result agreed with Jajhara et al., (2018) in their study about "A study on quality of life among thalassemic children aged 8 to 18 years ". They reported Thalassemia has a negative impact on perceived physical, emotional, social and school functioning in thalassemia patients. There was a significant improvement in the total quality of life with compliance with blood transfusion and regular iron chelation therapy.

Bedsides, this finding was in line with Dehnoalian et al., (2017) in their study about "The Impact of Educational Counseling Program on Quality of Life of Thalassemia Patients ". They reported that educational counseling programs improved the quality of life of patients with thalassemia.

In the same context, the study was consistent with El-Said Zaghamir al., (2019) in their study about "Impact of Educational Program about Iron Chelation Therapy on the Quality of Life for Thalassemic Children". This study revealed that there were statistically significant improvement of children's quality of life at follow up compared with pre-intervention phase.

Furthermore, this finding was in line with Bakhshi (2018) in their study about " The effect of group counseling on the quality of life in patients with major thalassemia referred to the thalassemia treatment center in Bushehr "They found that group counseling with cognitive behavioral approach significantly improves the quality of life in patients with Thalassemia major.

Moreover, this result came in agreement with Wang et al., (2022) in their study about "Psychological intervention in children with transfusion-dependent β-thalassaemia" .They concluded that Psychological intervention has positive effects on the treatment for children with transfusion-dependent β-thalassaemia, and it significantly improved their scores of QoL.

Also, this result agreed with Borhani et al., (2011) in their study about

"The effect of family-centered empowerment model on quality of life of school–aged children with thalassemia major". They reported that there was a statistically significant difference in quality of life of school–aged children with thalassemia after implementation of family-centered empowerment model.

To capitulate children had higher level of knowledge and self-care practices about thalassemia tremendously after implementation of health education. As well as, their quality of life was improved.

Conclusion

Based on the finding of the present study, the following is concluded:

Children with thalassemia who received self-care determination theory based intervention had higher level of knowledge about thalassemia on

posttest than pretest. Also, Children with thalassemia who received self-care determination theory based intervention had higher level of autonomy, relatedness and satisfaction on posttest than pretest .Meanwhile; they had higher quality of life on posttest than pretest.

Recommendations

In the light of the findings obtained from the current study and its conclusion, the following recommendations are suggested:

1. Self-determination theory based intervention about thalassemia should be designed and implemented in pediatric thalassemia units to improve children's knowledge and practices on the basis of their actual needs.
2. Self-determination theory based intervention about thalassemia should be designed and implemented in pediatric thalassemia units to improve parent's knowledge and practices to provide optimum care to children.

B. Recommendations for future Research: -

Further studies should be applied on a larger sample to determine the effect of self-determination theory based intervention to ensure the generalizability of results.

References

Abu Samra, O., Auda, W., Kamhawy, H., & Al-Tonbary, Y. (2015). Impact of educational programme regarding chelation therapy on the quality of life for B-thalassemia major children. Hematology, 20(5), 297-303.

Ansteenkiste, M., Delesie, L. M., Mariman, A. N., Soenens, B., Tobback, E., ... & Vogelaers, D. P. (2015). Examining the role of psychological need satisfaction in sleep: A Self-Determination Theory perspective. Personality and individual differences, 77, 199-204.

Atshan, R. S., & Aziz, A. R. (2022). Effectiveness of an Educational Program on Knowledge about Home Health Care Management to Children with Beta Thalassemia-Major at Thalassemia Center in Al-Zahra Teaching Hospital for Maternity and Children in Al-Najaf City. Pakistan Journal of Medical & Health Sciences, 16(03), 931-931.

Baghersalimi, A., Darbandi, B., Kazemnezhad Leyli, E., Kamran Mavardiani, Z., Ahmad Sharbafi, M., & Rezasefat Balesbaneh, A. (2021). Evaluation of self-efficacy in children and adolescents with thalassemia major. Journal of Pediatric Hematology/Oncology, 43(6), e754-e758.

Bakhshi, R., Bahreini, M., Mirzaei, K., & Kiani, J. (2018). The effect of group counseling on the quality of life in patients with major thalassemia referred to the thalassemia treatment center in Bushehr. Pajouhan Scientific Journal, 16(3), 11-19.

Benz Jr, E. J., & A Williams, G. C., McGregor, H. A., Zeldman, A., Freedman, Z. R., & Deci, E. L. (2014). Testing a self-determination theory process model for promoting glycemic control through diabetes self-management. Health Psychology, 23(1), 58.

Biswas A., Shawky R. and Kamal T. (2019): Thalassemia intermedia: An overview, Egyptian journal of medical

human genetics science direct:13(3) P. 245- 255.

Borhani, F., Najafi, M. K., Rabori, E. D., & Sabzevari, S. (2011). The effect of family-centered empowerment model on quality of life of school–aged children with thalassemia major. Iranian journal of nursing and midwifery research, 16(4), 292.

Dehnoalian, A., Madadkar Dehkordi, S., Alaviani, M., Motamedi, Z., Ahmadpour, S., & Banan-Sharifi, M. (2017). The Impact of Educational Counseling Program on Quality of Life of Thalassemia Patients. Jundishapur Journal of Chronic Disease Care, 6(4).

Elalfy, M. S., Farid, M. N., Labib, J. H., & RezkAllah, H. K. (2014). Quality of life of Egyptian b-thalassemia major children and adolescents. The Egyptian Journal of Haematology, 39(4), 222.

Eldakhakhny, A. M. M. (2011). Quality of life of school age thalassemic children at Zagazig City. Journal of American Science.Abolwafa, N. F., Mohamed, A. H., & Mohamed, A. A. (2019). Quality of Nursing Care among School Age Children with Thalassemia as Regards Blood Transfusion and Self Concept. American Journal of Nursing, 7(5), 670-676.

Eliezer A., Rachmilewitz . and Giardia P. (2016) : How I treat thalassemia: www.bloodjournal.org :61(3); P. 3479.

El-Kamah, G., Adam, S., Magdy, P., Michel, M., & Afifi, H. (2014). Quality of Life Outcomes Among Egyptian Children with Beta-Thalssemia. Blood, 124(21), 4864.

El-Said Zaghamir, D., Hanie El-Kazaz, R., Ahmed Khalil Morsy, A., & Ahmed Elmazahy, M. M. (2019). Impact of Educational Program about Iron Chelation Therapy on the Quality of Life for Thalassemic Children. Port Said Scientific Journal of Nursing, 6(3), 33-49.

Elsoudy, S. F., Madinay, N., & Yonis Mahrous, E. S. (2022). Compliance of Children with Thalassemia to Their Therapeutic Regimen. International Egyptian Journal of Nursing Sciences and Research, 2(2), 168-182.

Elzaree, F. A., Shehata, M. A., El Wakeel, M. A., El-Alameey, I. R., AbuShady, M. M., & Helal, S. I. (2018). Adaptive functioning and psychosocial problems in children with beta thalassemia major. Open Access Macedonian Journal of Medical Sciences, 6(12), 2337.

Gharaati, F., & Aghamolaei, T. (2019). Effect of a mobile-phone mediated based education on self-care behaviors of patients with thalassemia major. Journal of Caring Sciences, 8(3), 149.

Hakeem, G. L. A., Mousa, S. O., Moustafa, A. N., Mahgoob, M. H., & Hassan, E. E. (2018). Health-related quality of life in pediatric and adolescent patients with transfusion-dependent ß-thalassemia in upper Egypt (single center study). Health and quali ty of life outcomes, 16(1), 1-9.

Hamamy, H. A., & Al-Allawi, N. A. (2013). Epidemiological profile of common haemoglobinopathies in Arab countries. Journal of community genetics, 4(2), 147-167.

Hamdy, M., Draz, I. H., El Sayed, I. T., Ayyad, A. A. F., & Salemd, M. R. (2021). Assessment of Quality of Life among Beta-Thalassemia Major Patients Attending the Hematology Outpatient Clinics at Cairo University Hospital. Open Access Macedonian Journal of Medical Sciences, 9(E), 156-160.

Jajhara,I ., Choudhary. G., Singh. J., Chachan. V., Kumar.A. (2021) A study on quality of life among thalassemic children aged 8 to 18 years. Intrtnational Journal of Contemporary Pediatrics. Oct; 8(10):1667-1674.

Kadhim, K. A., Baldawi, K. H., & Lami, F. H. (2019). Prevalence, incidence, trend, and complications of thalassemia in Iraq. Hemoglobin, 41(3), 164-168.

Kahouei M., Kazemzadeh F., Zadeh J. and Ahmadi Z. (2016): Hierarchy of Iranian parents information needs and social seeking behavior of infants suffering blood disease: the social sciences:31(6); P. 336.

Kaur, R., Kaur, R., Kaur, S., Marwaha, L. R., & Bansal, D. (2015). Effectiveness of Cartoon Picture Book on self-care behavior of children between 4-10 years of age suffering from Thalassemia. Nursing & Midwifery Research Journal, 11(4), 185-197.

Madmoli, Y., Salimi, M., Madmoli, M., Maraghi, E., Pelarak, F., Korkini, N., & Mashalchi, H. (2019). The effect of orem self-care model on health-related quality of life of patients with thalassemia major. Journal of Research in Medical and Dental Science, 7(2), 170-176.

Mahmoud, M. M., Helaly, N. M., Mohammed, A. A. E. R., & Kamal, M. Y. (2019). Quality Of Life of Children with Thalassemia in Alexandria. Alexandria Scientific Nursing Journal, 21(1), 69-80.

Mazzone, L., Battaglia, L., Andreozzi, F., Romeo, M. A., & Mazzone, D. (2019). Emotional impact in β-thalassaemia major children following cognitive-behavioural family therapy and quality of life of caregiving mothers. Clinical practice and epidemiology in mental health, 5(1), 1-6.

Mediani, H., Hendrawati, S., & Susanti, D. (2019). Correlation Between Family Support and Medication Adherence of School-Age Thalassemic Patients at The Thalassemia Clinic in Indonesia. In Enhancing Capacity of Healthcare Scholars and professionals in Responding to the Global Health Issues (pp. 189-198). Sciendo.

Moghadam, M. P., Nourisancho, H., Shahdadi, H., Shahraki, S., Azarkish, B., & Balouchi, A. (2016). Effects of home-care training on the self-efficacy of patients with beta thalassemia major. Materia socio-medica, 28(5), 357.

Naggrawal A., Arorams A., Dewan P. and Harish K. (2011): Essential pediatric nursing: 3rd , India, CBS publishers α ,distributors PVT. LTD, P.P; 447, 448,449, 450, 451.

Navaneetha, K., S,, Shaji, G., Ravalya, P., & Nazi, S.K. (2013). Management of thalassemia. International research journal of pharmacy, 4(10), 1-3.

ngelucci, E. Diagnosis of thalassemia (adults and children), 2022.

Available at: https://www.wolterskluwer.com/en/know/clinical-effectiveness-terms

Nourbakhsh, S. M. K., Atamanesh, M., Effatpanah, M., Salehi, M., & Heidari, M. (2021). The Association between Behavioral Problems with Self-Esteem and Self-Concept in Pediatric Patients with Thalassemia. Iranian Journal of Psychiatry, 16(1), 36.

Ntoumanis, N., Ng, J. Y., Prestwich, A., Quested, E., Hancox, J. E., Thøgersen-Ntoumani, C., ... & Williams, G. C. (2021). A meta-analysis of self-determination theory-informed intervention studies in the health domain: effects on motivation, health behavior, physical, and psychological health. Health Psychology Review, 15(2), 214-244.

Potts N. and Mandleco B. (2012): Pediatric nursing caring for children and their families, 3rd ed. Australia: Delmar:12(1); P. 926.927.928.

Ragb, S. M., Elrahim, M. A., El Fotoh, W. M., & Ibrahim, R. A. (2021). Screening of β-thalassemia carriers in high school students in Shebin El-Kom, Menoufia Governorate. Menoufia Medical Journal, 34(1), 237.

Raza S., Ishfag K. & Fazal H. (2015): social impact of thalassemia major on patients families: 2(3); P.413.

Shackleford, J. (2018). Applying the Self-Determination Theory to Adolescents with Congenital Disease.

Safizadeh H., Farahmandinia Z., Soltaninejad S., Pourdanghan N. and Araste M. (2012): Quality of life in patient with thalassemia major and intermedia in kerman- iran : Mediterranean journal of hematology and infectious disease: 4(1) ; P.4.5.

Saha, R., Misra, R., & Saha, I. (2016). School functioning activity of Bengali thalassemic children attending a tertiary care hospital of eastern India. International Journal of Travel Medicine and Global Health, 4(3), 82-87.

Shanshory M., Hagag., sheble., AbdElhamed., Abd El- Bar., Al- Tonbary., Mansour., Hassan., Hamdy., Alfy., Sherief. and Sharaf, (2021) : spectrum of beta globin gene mutations in Egyptian children with B- thalassemia: Mediterranean journal of hematology and infectious disease:11(8); P. 2.

Wilson D. and Wang, M., Huang, M., & Hong, Y. (2022). Psychological intervention in children with transfusion-dependent β-thalassaemia. Vox Sanguinis, 117(3), 386-392.

Hockenberry M. (2011): Nursing care of infants and children, 9th, china, Elsevier; P.1436, 1437, 1438, 1439.