Quality of Life of School Age Thalassemic Children at Zagazig City

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Abstract: Background: The assessment of quality of life in children, especially in those with chronic illness such as Thalassaemia, is particularly important. It differs from other forms of medical assessment in that it focuses on the individuals' own views of their well-being and other aspects of life, giving a more holistic view of well-being. The aim of the present study was to: assess the quality of life of school-age children with Thalassemia at Zagazig City. Subjects And Methods: A descriptive study was conducted on a sample of 100 school-age thalassemic children at out-patient Hematology clinic at Zagazig University Hospitals in Sharkia Governorate, Egypt. Two tools were used to collect the necessary data. The first was a structured interview questionnaire sheet including socio-demographic data of children and their parents as well as medical history. The second tool was a standardized tool (the Pediatric Quality of Life Inventory ^{TM} Version 4.0). Results: The results of the present study revealed that the quality of life of school-age children with Thalassemia Major was affected. There was a significant association between the total quality of life and compliance with blood transfusion in both child and parent report. In addition, there was a significant association between the total quality of life and regular iron chelation therapy. Concluosion: Thalassaemia has a negative impact on perceived physical, emotional, social and school functioning in thalassaemia patients. Recommendations: Suitable programs aiming to increase children's adherence to the treatment regimen should be provided to increase psychosocial support.

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Key words: Quality of life, thalassemia major, school-age.

1. Introduction:

Thalassemias are inherited disorders of Hb synthesis that result from an alteration in the rate of globin chain production. A decrease in the rate of production of a certain globin chain or chains (α, β, γ, δ) impedes Hb synthesis and creates an imbalance with the other, normally produced globin chains (Madara & Pomarico-Denino, 2007).

The most severe form is the β-thalassemia major, which is characterized by a severe microcytic, hypochromic anemia (Cooley's anemia), whose symptoms appear usually within the first 2 years of life. Infants become pale and asthenic, have a poor appetite, grow slowly, and often develop jaundice; spleen, liver, and heart may also be enlarged. Adolescents with the most severe form may experience delayed puberty ((Mazzone et al, 2009)).

β-thalassemia is the most common chronic hemolytic anemia in Egypt (85.1%). A carrier rate of 9-10.2% has been estimated in 1000 normal random subjects from different geographical areas of Egypt (El-Beshlawy, 1999).

Children with thalassemia major have good survival but little is known about their quality of life (Sachdeva et al, 2002). Children not only have longer lives to be compared to adults, but they are less able to voice their concerns and are more vulnerable than adults (Ismail et al, 2006).

The assessment of quality of life (QOL) in children especially in children with chronic illness such as thalassaemia is particularly important (Ismail, 2006). An assessment of QOL differs from other forms of medical assessment in that it focuses on the individuals' own views of their well-being and assesses other aspects of life, giving a more holistic view of well-being (Dahlui et al., 2009).

It is important to understand more about quality of life in pediatric population to evaluate and improve the care patients receive. Children with chronic physical illness exemplified thalassemia are vulnerable to emotional and behavioral problems leading to poor quality of their lives. The disease may cause a sense of stigmatization in the child leading to feeling of shame and rejection. It also may affect social relations, school interactions, and self-esteem. So, great attention has to be taken especially by the nurse during follow up and treatment.

The aim of the present study was to

Assess quality of life of school age children with thalassemia.

2. Subjects and Methods:

Research design

A descriptive study was conducted to assess the quality of life of school-age children with thalassemia major.

Research Question

What is the impact of thalassemia as a chronic disease on the quality of life of school-age children?

I- Subjects

1. Setting

The study was conducted at Out-Patient Hematology Clinic at Zagazig University Hospitals in Sharkia Governorate.

2. Subject

The study was conducted on a sample of 100 school-age children who fulfilled the following criteria:-

1- Confirmed diagnosis of thalassemia major.
2- Age: from 6 years old to 12 years old
3- Both sexes.
4- Free from any other chronic diseases.

3. Tools for data collection

Two tools were used to collect the necessary data.

Tool I: Structured interview questionnaire sheet

Structured interview questionnaire sheet was developed by the researcher to collect the following necessary data:

A- Biosocial data of the child as, child's age, sex, birth date, and level of education.
B- Information about disease history, any similar conditions in the family, number of blood transfusions per month, compliance with blood transfusion and compliance with chelation therapy.
C- Socio-demographic data of child's parents, such as, age, educational level, occupation, family income as well as crowding index.

Tool Π: The Pediatric Quality of Life Inventory™ Version 4.0 by Varni et al., 1998.

The Peds QL™ Measurement Model is a modular approach to measure health-related quality of life (HRQOL) in healthy children and adolescents and those with acute and chronic health conditions. The Peds QL™ Measurement Model integrates seamlessly both generic core scales and disease-specific modules into one measurement system.

This form includes:

- Young children report (ages 5-7)
- Parent report for young children (ages 5-7)
- Child report (ages 8-12)
- Parent report for children (ages 8-12)

There are four domains in each report. The PedsQL version 4.0 consists of 23 items including the following:

1) Physical functioning (eight items)
2) Emotional functioning (five items)
3) Social functioning (five items)
4) School functioning (five items)

Scoring system for assessment of the quality of life (Varni et al., 1998):

The Peds QLTM 4.0 Generic Core Scales are comprised of parallel child self-report and parent proxy-report formats. Child self-report includes ages 5-7, and 8-12 years. Parent proxy-report includes ages 5-7 (young child), and 8-12 (child), and assesses parent's perceptions of their child's HRQOL. The instructions ask how much of a problem each item has been during the past 1 month.

0 = I never have a problem
1 = I almost never have a problem
2 = I sometimes have a problem
3 = I often have a problem
4 = I almost always have a problem.

Π- Methods

1- An official permission was obtained to facilitate collection of data.

2- Sociodemographic questionnaire sheet was developed by the researchers after thorough review of literature.

3- Jury was done to the tool by 5 experts (Three professors of pediatric nursing, one professor of hematology, and one professor of statistics).

4- The Pediatric Quality of Life InventoryTM Version 4.0 by Varni et al., 1998.

5- Written consent was obtained from parents of children.

6- A pilot study was conducted on 10% of children and their parents to test the clarity of questions and to estimate the time required for filling the sheet and no modification was done.

7- Each child and his/her mother were individually interviewed to collect the necessary data using tool I and tool Π. The time consumed to answer the questionnaire sheet ranged from 25 to 30 minutes. The average number of children and their mothers/day was 6 children.

8- Data was collected during 10 months, starting from July 2008 to April 2009.

Statistical analysis

The collected data was coded and entered in a data base file using the Foxpro for windows program. After complete entry, data were transferred to the SPSS version 14.0 program by which the analysis was conducted applying frequency tables with percentages and cross tabulations. The chi-square test was used to find the significant associations between the demographic and clinical data and the outcome measures.

3. Results

Table (1) shows the characteristics of the studied thalassemic children. Regarding the children's age, 24% were from 6 to 7 years. While 76% were aged from 8 to 12 years with mean age of 9.29±2.17 years. Also, 56% were males and 44% were females. Those who ranked the first birth order constituted 28%, the second 25%, while 24% were the third. Only 8% were either the fifth or more in birth order. It is revealed from the same table that 76% were from rural compared to 24% were from urban.

Regarding family income, 20% of the studied children's families had insufficient income, 69% had slightly sufficient income. On the other hand, 11% had sufficient and save income. As regard to crowding index, 37% of families of the studied children were with crowding index of < 2, while 55% were with crowding index of 2-3, as well as 8% only had crowding index of >3 .

Table (1): Characteristics of the Studied Thalassemic Children

Characteristics	No. (100)	%
Age in years:		
6-7 years	24	24.0
8-12 years	76	76.0
Mean ± SD	**9.29±2.17**	
Sex:		
Male	56	56.0
Female	44	44.0
Birth order:		
The first	28	28.0
The second	25	25.0
The third	24	24.0
The fourth	15	15.0
The fifth and more	8	8.0
Residence:		
Rural	77	77.0
Urban	23	23.0
Crowding index		
< 2	37	37.0
2-3	55	55.0
> 3	8	8.0
Family income :		
Insufficient	20	20.0
Slightly sufficient	69	69.0
Sufficient and save	11	11.0

Table (2) shows the medical history of the studied thalassemic children. It reveals that children who were diagnosed as β-thalassemia major by the first year of life constituted 83%, while 17% only were diagnosed by the second year. Consanguinity was found among 65% of the parents, while 35% of them negative consanguinity. It was also found that 57% had similar conditions in the family and 43% had no conditions.

Regarding compliance with treatment, 48% of the studied children had irregular blood transfusion therapy, while 52% were regulars. In addition, 42% of the studied children were compliant with iron chlation therapy, while 58% were not compliant.

It was revealed from the same table that, 68% of the studied children come to the hospital once per month for transfusion therapy, while 32% come twice per month.

Table 3 and figure 1 portray the total QOL score and its domains. According to child's report, 37% of children had good score compared to 21% in parent's report, while 58% had fair score compared to 64% in parent's report.

Table (2): Medical History of the Studied Thalassemic Children

Medical History	No. (100)	%
Disease occurrence:		
By 1st year	83	83.0
By 2nd year	17	17.0
Consanguinity:		
Present	65	65.0
Not present	35	35.0
Similar conditions in the family:		
Present	57	57.0
Not present	43	43.0
Transfusion therapy:		
Regular	52	52.0
Irregular	48	48.0
Iron chelation therapy:		
Regular	42	42.0
Irregular	58	58.0
No. of transfusion therapy/month:		
Once	68	68.0
Twice	32	32.0

Table (3): The Total QOL Scale and QOL Subscales of the Studied Thalassemic Children

QOL scale	*Very good* =100%		Good 75-99.9		Fair 50-74.9		Bad 25-49.9		Very bad 0-24.9		Mean ± SD
	N	*%*	*N*	*%*	*N*	*%*	*N*	*%*	*N*	*%*	
Child report:											
Total QOL	0	0.0	37	37.0	58	58.0	5	5.0	0	0.0	73.1±12.4
Physical	3	3.0	56	56.0	39	39.0	2	2.0	0	0.0	63.1±17.8
Emotional	1	1.0	28	28.0	47	47.0	21	21.0	3	3.0	60.2±20.1
Social	5	5.0	57	57.0	30	30.0	7	7.0	1	1.0	84.15±12.4
School	2	2.0	35	35.0	46	46.0	17	17.0	0	0.0	74.95±16.5
Parent's report:											
Total QOL	0	0.0	21	21.0	64	64.0	15	15.0	0	0.0	64.8±13.8
Physical	1	1.0	25	25.0	48	48.0	22	22.0	4	4.0	60.62±19.7
Emotional	0	0.0	23	23.0	43	43.0	27	27.0	7	7.0	54.8±21.14
Social	4	4.0	57	57.0	28	28.0	10	10.0	1	1.0	73.05±17.8
School	4	4.0	33	33.0	47	47.0	15	15.0	1	1.0	66.55±17.1

Regarding physical QOL, 56% had good score according to child's report compared to 25% in parent's report, while 39% had fair score compared to 48% in parent's report.

In relation to emotional QOL, 28% had good score according to child's report compared to 23% in parent's report. On the other hand, 21% had bad score compared to 27% in parent's report, while 47% had fair score compared to 43% in parent's report.

Regarding social QOL, 57% had good social score in both child and parent's report, while 30% had fair social score compared to 28% in parent's report.

Regarding school QOL, 35% had good score according to child's report compared to 33% in parent's report. Forty six percent of children had fair score compared to 47% in parent's report while, 17% of children had bad score compared to 15% in parent's report.

The same table reveals that the emotional functioning scored the lowest followed by physical then school and social functioning according to both child and parent's report.

The QOL scores of the studied thalassemic children regarding the physical and emotional functions are illustrated in table 4. In the child's report, it was found that 79% of children had very good score regarding taking a bath alone compared to 76% in parent's report. As regard walking, according to child's report, 66% had very good score compared to 57% in parent's report.

Table (4): Distribution of the Physical and emotional Functioning of the Studied Thalassemic children

Physical and Emotional domains	Very good		Good		Fair		Bad		Very bad		Total
	N	%	N	%	N	%	N	%	N	%	
Physical domain											
Child report:											
Walking	66	66.0	1	1.0	26	26.0	2	2.0	5	5.0	100
Running	18	18.0	0	0.0	25	25.0	12	12.0	45	45.0	100
Making activity	32	32.0	3	3.0	44	44.0	11	11.0	10	10.0	100
Lifting something	43	43.0	7	7.0	28	28.0	13	13.0	9	9.0	100
Taking a bath	79	79.0	0	0.0	12	12.0	4	4.0	5	5.0	100
Doing chores	51	51.0	12	12.0	28	28.0	6	6.0	3	3.0	100
No pain	31	31.0	0	0.0	49	49.0	14	14.0	6	6.0	100
Having energy	20	20.0	3	3.0	39	39.0	18	18.0	20	20.0	100
Parent report:											
Walking	57	57.0	5	5.0	25	25.0	9	9.0	4	4.0	100
Running	17	17.0	1	1.0	28	28.0	17	17.0	37	37.0	100
Making activity	36	36.0	4	4.0	41	41.0	10	10.0	9	9.0	100
Lifting something	38	38.0	4	4.0	34	34.0	14	14.0	10	10.0	100
Taking a bath	76	76.0	4	4.0	11	11.0	4	4.0	5	5.0	100
Doing chores	46	46.0	11	11.0	32	32.0	7	7.0	4	4.0	100
No pain	21	21.0	1	1.0	51	51.0	22	22.0	5	5.0	100
Having energy	16	16.0	3	3.0	30	30.0	26	26.0	25	25.0	100
Emotional domain											
Child report:											
Being afraid	46	46.0	2	2.0	23	23.0	7	7.0	22	22.0	100
Being sad	38	38.0	3	3.0	44	44.0	9	9.0	6	6.0	100
Being angry	14	14.0	0	0.0	22	22.0	16	16.0	48	48.0	100
sleeping trouble	47	47.0	2	2.0	31	31.0	13	13.0	7	7.0	100
Being worried	65	65.0	6	6.0	15	15.0	10	10.0	4	4.0	100
Parent's report:											
Being afraid	44	44.0	2	2.0	18	18.0	13	13.0	23	23.0	100
Being sad	37	37.0	2	2.0	34	34.0	16	16.0	11	11.0	100
Being angry	8	8.0	1	1.0	14	14.0	19	19.0	58	58.0	100
Sleeping trouble	42	42.0	5	5.0	29	29.0	17	17.0	7	7.0	100
Being worried	56	56.0	6	6.0	16	16.0	13	13.0	9	9.0	100

On the other hand, 45% of the studied thalassemic children had bad score regarding running and no one had very bad score according to child's report, while in parent's report, 17 % had bad score and 37% had very bad score.

Regarding having energy, 20% had very bad score according to child's report compared to 25% in parent's report. Thirty nine percent of children had fair score compared to 30% in parent's report.

As regard activities and exercises, according to child's report, 32% had very good score and 44% had fair score. In comparison with parent's report, 36% had very good score and 41% had fair score.

According to child's report, 43% had very good score regarding lifting something heavy and 9% had very bad score, while in parent's report 38% had very good score and 10% had very bad score .

Regarding pain, 49% had fair score and 14% had bad score according to child's report. While in

parent's report, 51% had fair score and 22% had bad score.

Among studied thalassemic children, 51% had very good score regarding doing chores around the house in child's report compared to 46% in parent's report and 28% had fair score compared to 32% in parent's report.

According to child's report, it was found that 46% of children had very good score regarding fear compared to 44% in parent's report. Those who had very bad score constituted 22% compared to 23% in parent's report.

Regarding sadness, 44% of children had fair score compared to 34% in parent's report. In addition, 38% had very good score compared to 37% in parent's report.

As regard anger, 48% of children had very bad score compared to 58% in parent's report. Only 14% had very good score compared to 8% in parent's report.

In relation to having sleeping trouble, 47% of children had no sleeping trouble and had very good score compared to 42% in parent's report. Children who had fair score constituted 31% compared to 29% in parent's report.

Regarding worry, 65% of the studied children had very good score as they had no worry about what will happen to them compared to 56% in parent's report. Ten percent of children had bad score compared to 13% in parent's report.

Table 5 illustrated the social and school functioning of the studied thalassemic children. According to child's report, it was found that 90% had no problems with getting along with other kids so they had very good score compared to 85% in parent's report. Children who had very good score regarding acceptance of them from other kids as friends constituted 72% compared to 75% in parent's report.

Table (5):Distribution of the Social and School Functioning of the Studied Thalassemic Children

Social and School domains	Very good		Good		Fair		Bad		Very bad		Total
	N	%	N	%	N	%	N	%	N	%	
Social Functioning											
Child's report:											
Getting along with kids	90	90.0	0	0.0	2	2.0	4	4.0	4	4.0	100
Other kids refuse him	72	72.0	0	0.0	20	20.0	4	4.0	4	4.0	100
Teasing from other kids	79	79.0	0	0.0	11	11.0	5	5.0	5	5.0	100
Can't do things as others	41	41.0	3	3.0	44	44.0	3	3.0	9	9.0	100
Keeping up when play	29	29.0	1	1.0	27	27.0	19	19.0	24	24.0	100
Parent report:											
Getting along with kids	85	85.0	0	0.0	5	5.0	4	4.0	6	6.0	100
Other kids refuse him	75	75.0	0	0.0	16	16.0	5	5.0	4	4.0	100
Teasing from other kids	74	74.0	0	0.0	14	14.0	9	9.0	3	3.0	100
Can't do things as others	38	38.0	4	4.0	42	42.0	6	6.0	10	10.0	100
Keeping up when play	26	26.0	2	2.0	26	26.0	21	21.0	25	25.0	100
School Functioning											
Child's report:											
Pay attention in class	58	58.0	3	3.0	29	29.0	7	7.0	3	3.0	100
Forgetting things	71	71.0	1	1.0	21	21.0	5	5.0	2	2.0	100
School work	84	84.0	1	1.0	13	13.0	1	1.0	1	1.0	100
Missing school due to illness	48	48.0	4	4.0	23	23.0	18	18.0	7	7.0	100
Missing school to go to hospital	5	5.0	1	1.0	13	13.0	8	8.0	73	73.0	100
Parent's report:											
Pay attention in class	60	60.0	3	3.0	25	25.0	9	9.0	3	3.0	100
Forgetting things	72	72.0	2	2.0	16	16.0	7	7.0	3	3.0	100
School work	85	85.0	1	1.0	13	13.0	1	1.0	0	0.0	100
Missing school due to illness	46	46.0	5	5.0	19	19.0	23	23.0	7	7.0	100
Missing school to go to hospital	7	7.0	0	0.0	12	12.0	8	8.0	73	73.0	100

Children who were not teased from others and had very good score constituted 79% that compared to 74% in parent report.

In relation to the ability of doing things as other kids, 41% had very good score compared to

38% in parent's report. Children had fair score rated 44% compared to 44% in parent's report.

Regarding keeping up with other kids when playing, 29% of children had very good score compared to 26% in parent's report, while 24% of children had very bad score compared to 25% in parent's report.

Regarding paying attention in class, according to children report it was shown that 58% of children had very good score compared to 60% in parent's report. Children who had fair score constituted 29% compared to 25% in parent's report.

As regard to forgetting things, 72% of children had very good score and the same percent was in parent's report. Regarding keeping up with schoolwork, 84% of children had very good score compared to 85% in parent's report.

In relation to missing school due to illness, 48% of children had very good score compared to 46% in parent's report and 18% had bad score compared to 23% in parent's report. Regarding missing school to go to hospital, 73% of children had very bad score, which was similar to parent's report.

The relation between the total QOL of the studied thalassemic children and number of blood transfusion per month is illustrated in table (6). It was found that 68% of children had blood transfusion once per month, while 32% had blood transfusion twice per month.

Table (6) Relation between the Total QOL of the Studied Thalassemic Children and Number of Blood Transfusion per Month

QOL scale	No. of blood transfusion / month				X^2	P-value
	Once/ month n = 68		*Twice/month* n = 32			
	No.	%	No.	%		
Child's report:						
Good	22	32.4	15	46.9	2.09	.35
Fair	42	61.8	16	50		
Bad	4	5.8	1	3.1		
Parent's report:						
Good	13	19.1	8	25.0	.57	.75
Fair	44	64.7	20	62.5		
Bad	11	16.2	4	12.5		

According to child's report, 46.9% of children who had blood transfusion twice per month had good QOL compared to 32.4% of children who had blood transfusion once per month. Children who had fair score in relation to having blood transfusion once per month constituted 61.8% compared to 50% of children who had twice per month.

According to parent's report, 25% of children who had blood transfusion twice per month had good score compared to 19.1% of children who had blood transfusion once per month. Children who had blood transfusion once per month and had fair score amounted up to 64.7% compared to 62.5% of children who had twice per month and had fair score.

This table shows no statistical significant difference in both child and parent's report between the total QOL and number of blood transfusion per month.

The relation between the total QOL of the studied thalassemic children and their compliance with blood transfusion therapy is illustrated in table (7). It was found that 52% of children were compliant with blood transfusion therapy, while 48% were not compliant.

Table (7) Relation between the Total QOL of the Studied Thalassemic Children and Compliance with Blood Transfusion

QOL scale	Compliance with blood transfusion				X^2	P-value
	Yes n = 52		No n = 48			
	No.	%	No.	%		
Child's report:						
Good	27	51.9	10	20.8	11.19	.004*
Fair	24	46.2	34	70.8		
Bad	1	1.9	4	8.3		

Parent's report:						
Good	16	30.8	5	10.4	7.3	.03*
Fair	31	59.6	33	68.8		
Bad	5	9.6	10	20.8		

According to child's report, 51.9% of compliant children had good QOL compared to 20.8% of children who were not compliant. Among compliant children 46.2% had fair score compared to 70.8% of not compliant children. On the other hand, 2.4% of compliant children had bad QOL compared to 6.4% of not compliant children

According to parent's report, 30.8% of compliant children had good QOL compared to 10.45% of not compliant children. Compliant children who had fair score constituted 59.6% compared to 68.8% of not compliant children and 9.6% of compliant children had bad QOL compared to 20.8% of not compliant children.

The table revealed that there was statistical significance between the total QOL of the studied thalassemic children and compliance with blood transfusion according to both child and parent's report (P value is statistically significant at < 0.05).

The relation between the total QOL of the studied thalassemic children and compliance with chelation therapy is illustrated in table (8). It was found that 41% of children were compliant with chelation therapy, while 59% were not compliant.

Table (8) Relation between the Total QOL of the Studied Thalassemic Children and Compliance with Chelation Therapy

QOL scale	Compliance with chelation therapy				X^2	P-value
	Yes n = 41		No n = 59			
	No.	%	No.	%		
Child's report:						
Good	22	53.7	15	25.4	8.5	.014*
Fair	18	43.9	40	67.8		
Bad	1	2.4	4	6.8		
Parent's report:						
Good	15	36.6	6	10.2	10.5	.005*
Fair	22	53.7	42	71.2		
Bad	4	9.8	11	18.6		

According to child's report, 53.7% of compliant children had good QOL compared to 25.4% of children who were not compliant. Compliant children who had fair score constituted 43.9% compared to 67.8% of not compliant children, while 2.4% of compliant children had bad QOL compared to 6.8% of non-compliant children

According to parent's report, 36.6% of compliant children had good QOL compared to 10.2% of not compliant children. About 53.7% of compliant children had fair score compared to 71.2% of not compliant children. On contrary, 9.8% of compliant children had bad QOL compared to 18.6% of not compliant children

In addition, this table also reveals that there was statistical significance between the total QOL of the studied thalassemic children and compliance with chelation therapy in both child and parent's report (P value is statistically significant at < 0.05).

4. Discussion:

The present study showed that more than half of the studied children were males. This finding is in consistent with the findings of Elsaid (2009) who conducted his study at Zagazig university Hospitals, Egypt and Salama et al (2006) who did his study at Mansoura University Children's Hospital, Egypt. The present study is also supported by Shaligram et al (2007) and Akbar (2004) who done his study in Shiraz city.

Similar to Shaligram et al (2007), the present study showed that more than three quarters (77%) of children were from rural area compared to 23% were from urban area. The findings of the present study are in contrast with Gharaibeh et al (2009) who did his study at the National Thalassemic Center in Damascus, Syria. Gharaibeh's study illustrated that about three quadrants were from urban. This reflects that thalassemia may be present in urban as in rural when premarital screening and counseling is neglected.

The current study found that most families reported insufficient family income. This finding goes in line with the findings of Gharaibeh et al (2009). This may explain that only poor families are more likely to take their children to general hospitals,

or may reflect that rich families have enough money to make premarital and prenatal screening tests when they are knowledgeable about it.

Akbar (2004) noticed that about half of cases were outcomes of first-or second-cousin marriages. This goes in line with the present study, which revealed that more than half of parents of the studied children were relatives. This may be due to strong family relationships in Egypt, especially those who live in rural areas. In addition, educational programs about genetic counseling are still neglected.

Gharaibeh et al (2009) found that more than two thirds of children (65.3%) had sick relatives with thalassemia. This goes in line with the present study. This may be as result of increased consanguineous marriages.

The present study found that more than half of children had no compliance with iron-chelation therapy. This finding is supported by Elsaid (2009) who done his study at Zagazig university Hospitals, and he found that most of his studied sample were not compliant with iron-chelation therapy. This may be due to the painful insertion of the needle subcutaneously, long periods of infusion or limited activity during the use of desferal bump. In addition to side effects of oral chelators which include abdominal pain, diarrhea and vomiting.

Regarding the total QOL of children with thalassemia major, Shaligram et al (2007) found that three quadrants of the studied thalassemic children had poor QOL. This goes in line with the present study as more than half of children had fair QOL and 5% had bad QOL.

The present study noticed that mothers related QOL of their children as fair in more than two thirds of them and bad in 15%. This finding was supported by Shaligram et al (2007) who mentioned that caregivers related QOL of their children as poor in most children. This was due to sense of guilt toward those children as thalassemia is an inherited disease and due to the permanent comparison between them and the other healthy children or siblings .

The current study found that emotional functioning scored the lowest followed by physical then school and social functioning. This finding is in consistent with Cheuk's et al (2008) study that conducted in Hong Kong. On other hand our finding is in contrast with both Thavorncharoensap et al (2010) and Ismail et al (2006) who found that school functioning scored the lowest, followed by emotional functioning and social functioning then physical functioning. Ismail and Thavorncharoensap et al (2010) clarified their finding by the fact that frequent absenteeism from school for hospital visits, and lack of energy when performing academic activities, had a significant negative impact on the children's health related quality of life (HRQOL).

Regarding physical QOL, Shaligram et al (2007) found that the majority of children had no problems with self-care followed by usual activities, and mobility. This finding is in agreement with the present study as 79% had no problems with self care, and more than two thirds had no problems with walking. Dahlui et al (2009) clarified that, the physical function scores were higher than the other domains because these patients had been having the disease since childhood, they were not working for a living and as such had not much expectation with regard to physical performance.

The present study found that slightly less than half had bad physical QOL regarding running while, one quadrant had bad physical QOL regard making activities and exercises. These findings are supported by Caro (2002) who found, less than one quadrant of conventionally treated thalassemia major patients had their activities very often stopped due to thalassemia, its complications or desferrioxamine treatment, and 20% had their physical activities limited at least a bit. This may be due to the regular period of mild anemia before the scheduled transfusion which might limit their exercise capacity as thalassemia leads to low hemoglobin level resulting in fatigue and general weakness (Cheuk et al, 2008).

The present study found that nearly half of children had affected QOL regarding pain and less than one quadrant had bad and very bad QOL. This finding is supported by Shaligram et al (2007) who clarified his result on the basis that iron chelating therapy produce arthritis, abdominal pain, diarrhea and vomiting which may have a bearing on the high score on the pain. In addition, Telfar et al (2005) mentioned that the fully-chelated patients had a QOL almost similar to that of normal children except with regard to body pain as he clarified that was because of complications and side effects of thalassemia treatment.

Regarding emotional QOL, the present study found that more than one third of children had fair emotional QOL and only 21% had bad emotional QOL. These findings are in agreement with a subsequent multi-center European study conducted by Sadowski et al (2002) found the same results. In addition, Pradhan et al (2003) found that more than two thirds of children had emotional problems.

Emotional QOL was affected because thalassemic children feel different from their peers and elaborate negative thoughts about their life. Children at this age are becoming more aware of themselves as individuals." They work hard at "being responsible, being good and doing it right." They are

now more reasonable to share and cooperate but the disease prevents them from being industry and the sense of inferiority may develop instead (Thavorncharoensap M, et al.2010).

In addition, children may develop psychological and emotional problems early from the toddler stage. Toddlers want to become capable of satisfying some of their own needs to develop a sense of autonomy but, if caregivers refuse to let them perform these tasks due to their illness, they may develop shame and doubt about their ability to handle problems. Moreover, the treatment is emotionally demanding, as transfusion and chelation therapy require repeated invasive procedures and hospital visits.

Regarding social functioning, Gharaibeh et al (2009) reported that stigmatization was significantly noticed among older children to younger children. He also found that all children who were not currently at school experienced severe stigmatization due to thalassemia major. While among those who were currently at school, 10.2% experienced a severe level of stigmatization. This explores that the young children who were at age of 6-12 years didn't feel disease stigma so, the majority of those children had very good level of social interaction with peers and school friends.

In consistent with Gharaibeh findings, the present study found that 79% of children, who had no teasing from others, had very good social QOL. Only 10% of children, who had teasing from others, had severe problems with social interaction.

In addition, the majority of children had no problems with getting along with other kids and they related their social QOL as very good. Regarding being a member in a play team with other kids, about three quadrants of the studied thalassemic children had no problems as other kids didn't refuse them so; they related their social QOL as very good. This may be due to the nearly absence of the disease complications at this age resulting in decreased feeling of stigmatization that causes limitations of social interaction. Also, (Musallam et al, 2008) mentioned that having an adjusted family has great benefits for thalassemic children as this enhances the child confidence and ability of being socially accepted.

Gharaibeh et al (2009) clarified that the difficulty of social interactions was reported less among children with thalassemia and this might be related to strong family relationships in Arabic culture.

Regarding school functioning, Saeed (2004) explored that having to go to hospital for blood transfusion and missing school is one of the most important factors affecting the QOL of conventionally treated thalassemic patients. In addition, Gharaibeh et al (2009) reported that education was one of the greatest difficulties that affected children with thalassemia, as 42.7% of children with thalassemia experienced moderate to severe difficulties in their education. Similarly, Cantaan et al. (2003) mentioned that education of two thirds of children with thalassemia at school age was affected, mainly due to having attended hospital for investigations and transfusions. Consistent with the previous studies, the present study found that slightly less than half had fair school QOL while, only 17% were bad.

Thavorncharoensap et al (2010) found no significant relationship between frequency of blood transfusion per year and health related quality of life (HRQOL). This finding is in agreement with the current study. Schrier (2004) clarified that regular transfusions should be started when the hemoglobin concentration falls below 7 gm/dL or when there is impaired growth because those patients who are able to maintain a hemoglobin concentration more than 7.5 gm/dL usually don't require chronic transfusion therapy. This may reflect that frequency of blood transfusion per month is associated with the pre-transfusion hemoglobin level and the severity of the disease.

Salama et al (2006) discussed that life expectancy of patients with thalassemia has greatly improved over the last decade because of regular transfusions and increased compliance with iron chelation therapy. In agreement with the current study, it was found that more than half of children who were compliant with blood transfusion therapy had good QOL compared to less than one quadrant of children who were non-compliant. This may be clarified by Dubey et al. (2008) who mentioned that regular blood transfusion promotes normal growth and physical activity, suppress erythropoisess and prevent chronic hypoxia and early splenomegaly and/or hypersplenism. Therefore, it is now possible for a thalassemic child to have a near normal life span with a good quality of life.

Telfar et al (2005), discussed that QOL for non-chelated and fully chelated thalassemia patients differed. The fully chelated patients had a QOL almost similar to that of normal children. In consistency, the present study revealed that more than half of children who were compliant with iron chelation therapy had good QOL and more than one third had fair score. On other hand, one quadrant only of non-compliant children had good QOL and more than half had fair score. This may be clarified by the fact that chelation therapy can reduce complications, and improve survival and quality of life of transfused patients (*Cianciulli*, 2009).

The current study revealed that more than half of children who were not compliant with iron chelation therapy had fair QOL and the minority of both compliant and not compliant children had bad QOL. These results may indicate that non compliance with chelation therapy had minimal effect on QOL of school-age thalassemic children. From the researcher point of view, this is not a fact because children did not leave the treatment completely; they only escaped for one or two days per week from the chelation regimen so, they had fair but not bad QOL. Only children who either completely escaped from chelation or chelation was neglected by parents had bad QOL.

5. Conclusion:

Based upon the results of the present study, it was concluded that thalassemia as a chronic disease had a negative impact on perceived physical, emotional, social and school functioning of school-age children resulting in impaired quality of life. The study reflected that emotional functioning was the most affected from the point of view of both children and parents. The study also demonstrated an association between compliance with treatment regimen and quality of life.

Based upon the finding of the current study, it may be recommended that:

- A psychologist in the hematological units and out patient clinics is a must be present to help in providing a link between patients, school officials, the families, and the physicians.
- An age-appropriate cartoon illustration for children including detailed scientific information about the disease and its survival should be provided.
- Mass media should have a role in providing information to population about Thalassemia and other inherited diseases (may be genetic counseling), and both discourage and highlight the disadvantages of consanguineous marriage.

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