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Assess The Quality of Life and Epidemiological Aspect of Pediatric with Sickle Cell Disease at Zagazig University Hospital

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ABSTRACT

Background: Sickle cell disease is considered one of the most common inherited diseases in the world. Quality of life is a broad multidimensional concept that usually includes subjective evaluations of both positive and negative aspects of life. **Objective:** To assess the quality of life and epidemiological aspect of pediatrics with sickle cell disease at Zagazig University Hospital. **Patients and Methods:** A cross-sectional study was conducted in the Outpatient Clinic and Pediatric Hematological Department in Zagazig University Hospital including 30 children with sickle cell anemia. We used two tools, the first was: Structure interview questionnaire sheets, and the second: was the Pediatric Quality of Life Inventory that was used to assess children's quality of life.

Results: The mean age of SCD patients is 10.65±3.41years.Our study showed that males (66.7%). In our study, we used the WHOQOL-BREF criteria for measuring QOL in SCD and found that children with SCD scored positive 76.7% in physical activity, 50% in emotional aspect, 63.3% in the social aspect, 46.4% in school aspect, and the total QOL assessment is 63.3%. Concerning the school achievements of children with SCD, the results of this study revealed that 53.3% of students didn't pay attention in class and 66.7% of the children forget things. There were statistically significant differences in all school health subitems.

Conclusion: It was concluded that sickle cell disease is a chronic disease that had a negative impact on QOL that includes physical, emotional, social, and school functioning. Our results reflected that all aspects of QOL were affected especially school functioning.

Keywords: Sickle cell disease; Quality of life; Epidemiology.

INTRODUCTION:

Sickle cell disease (SCD) is a group of blood disorders typically inherited. The most common type is known as sickle cell anemia (SCA). It results in an abnormality in the oxygen-carrying protein hemoglobin found in cells. This leads to a rigid, sickle-like shape under certain circumstances. Problems in sickle cell disease typically begin around 5 to 6 months of age [(1)].

Several health problems may develop, such as attacks of pain ("sickle cell crisis"), anemia, swelling in the hands and feet, bacterial infections, and stroke. Long-term pain may develop as people get older. The average life expectancy in the developed world is 40 to 60 years [(2)].

Sickle cell disease occurs when a person inherits two abnormal copies of the hemoglobin gene, one from each parent. This gene occurs in chromosome11. Several subtypes exist, depending on the exact mutation in each hemoglobin gene [(3)].

Quality of life is a broad multidimensional concept that usually includes subjective evaluations of both positive and negative aspects of life [(4)].

The study aimed to assess the quality of life and epidemiological aspect of pediatrics with sickle cell disease at Zagazig University Hospital.

PATIENTS AND METHODS

This study was a cross-sectional study conducted in outpatient Clinic and Pediatric Hematological Department in Zagazig University Hospital Including 30 children with sickle cell anemia and their parents who fulfill the following criteria, A- age ranges from 5 to18 years. B- free from any associated diseases. We used two tools, the first was: Structure interview questionnaire sheets, and the second: was the Pediatric Quality of Life Inventory that was used to assess children's quality of life.

Tools of data collection: Two tools were used to collect the necessary data for this study.

Tool I: Structure interview questionnaire sheets.

It was included the demographic data of children; age, gender, level of education, birth order, and residence. Parents demographic data; age, level of education, occupation, and income. The children's clinical data comprised the onset, duration, and type of treatment of sickle cell diseases such as blood transfusion: frequency and side effect of blood transfusion, and compliance with chelating therapy.

Tool II: Pediatric Quality of Life Inventory TM Version 4.0 by Varni *et al.* [(5)]

The Ped QLTM Measurement model is a modular approach to measure health-related quality of life 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 (A) Report for young children (age 5-7). (B) Parent Report for young Children (age5-7).

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(C) Report for young Children (age 8-18), and (D) Parent Report for young Children (age8-18).

There are four domains in each report. The Peds QL version 4.0 consists of including the following: (1) Physical functioning. (2) Emotional functioning. (3) Social functioning. (4) School functioning.

Physical function of children related to the disease process including:

- Physical activities, such as feeling tired during home activities, personal hygiene, bathing, running of short distance, walking of long distance, performing exercise and avoiding physical activities in cold weather.
- Dietary habits as adequate intake of iron, folic acid, vitamin C protein, copper and cobalt, water, water with fever, and exercise. Avoiding iron and vitamin C intake with frequent blood transfusion and avoiding taking cold drinks.
- Rest and sleeping patterns such as enough sleeping during the night, sleeping interruption during night related to disease, taking enough rest at daytime, and taking a warm bath when physically tired
- **School functioning:** that includes paying attention in class, keeping up with schoolwork, forgetting things, frequently being absent from school due to not feeling well as well as going to the doctor or hospital.
- **Emotional functioning:** it includes the following: feeling afraid or scared, feeling depressed and sad, feeling angry and worrying about what will happen to him or her, and difficulty in communicating with others like telling others when to feel pain and to what extent feel pain.
- **Social functioning:** it is measured by the following: getting about with other children, other children not wanting to be his or her friend, getting teased by other children, not being able to do things that children in the same age can do, and keeping up with other children.
- Children's dealing approach with the disease symptoms such as using comfortable measures stay in a calm place and keeping away from light, taking some drinks as Ginger, Mint, Lemon, and taking of analgesic in case of headache, using wet cloth (tipped compresses), hospital admission or taking medication in fever, avoiding of tight clothing and cold application in arms and legs swelling, and using non-pharmacological agents as a distraction, relaxation and massage and taking medication according to doctors order in bone and joint pain.
- The experienced pain intensity as mild, moderate, and severe, sites of feeling pain as the whole body, arms, legs, chest, back, and stomach.

Scoring system for assessment of the quality of life [5]:

The Peds TM4.0 Generic Core Scales are comprised of parallel children self-report and parent proxy-report formats. The instructions ask how much 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.

Items are reverse scored linearly transformed to a 0-100 scale as following: 0=100 (quality of life is very good=not affected QOL). 1=75(quality of life is good=mildly affected QOL). 2=50 (quality of life is fair=moderately affected QOL). 3=25 (quality of life is bad=severely affected QOL). 4=0 (quality of life is very bad=severely affected QOL). This means that the higher the score indicate better HRQOL.

All patients were subjected to the following:
A-Complete history taking
B-Thorough clinical examination.
C-Laboratory investigation: Complete blood count. (CBC), serum ferritin level, and hemoglobin Electrophoresis.
D- Specific investigation:

Every child and his mother were interviewed individually for 30-40 minutes to collect the necessary data .The researcher introduced herself and explained the purpose of the study briefly to them.

Collect data using; Tool 1; Structure interview questionnaire sheets. And Tool II: Pediatric Quality of Life. Of all subjected patient and their parents. Data were collected for 6 months from September 2019 to March 2020.

Ethical consent:
Approval of the study was obtained from Zagazig University academic and ethical committee. Informed written consent was obtained from parents of all children participants before recruitment in the study, after explaining the objectives of the work. This work has been carried out following The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

Statistical Analysis

The collected data were coded, processed, and analyzed using the SPSS (Statistical Package for Social Sciences) version 22 for Windows® (IBM SPSS Inc, Chicago, IL, USA). Data were tested for normal distribution using the Shapiro Walk test. Qualitative data were represented as frequencies and relative percentages. Chi-square test ($\chi 2$) to calculate the difference between two or more groups of qualitative variables. Quantitative data were expressed as mean ± SD (Standard deviation). Independent samples t-test was used to compare between two independent groups

of normally distributed variables (parametric data). P-value < 0.05 was considered significant.

RESULTS

Table (1) and (2) illustrate; the mean age of studied children10.65±3.41years, 66.7% of them were males Concerning types of SCD, 66.7% is SCA.

Table (3) shows; 23.3% were poor regard physical aspect, 50% were poor regard emotional aspect, 36.7% were poor regard social aspect, 53.3% were poor regard school aspect and overall QOL 36.7% were poor and 63.3% were good

This table shows; 73.3% of children had a very good score in walking, 66,7% of children had a very good score in making activity, 53.3% had a very good score in running, 83.3% had a very good score in taking a bath, while only 26.6% of studied children had a very good score in having energy and 76.7% had a very good score regarding doing chores around the house. Regarding pain, 43.3% had a very good score.

50% had a very good score regarding fear. Regarding sadness, 46.7% had a very good score. As regard anger, 23.3 % had a very good score. Concerning sleeping trouble, 10% had a very bad score. About worry, 16.7% of the studied children had a very good score.Children who were not teased by others had a very good score of 93.3% which was the same result about acceptance of children from other kids as friends. It was found that 90% had no problems with getting along with other kids.

Regarding keeping up with other kids when playing, 66.7% of children had a very good score. Concerning the ability to do things as other kids 60% had a very good score.

23.3%of the studied sickle cell children had a very good score regarding paying attention in class, as regard to missing school due to illness 50% had a very bad score. Also missing school to go to hospital 53.3% of children had very bad score. As regards forgetting things, 40% of children had bad scores **Table (4)**.

Table 5 illustrates the total QOL score and its domains. 30% of children had good scores while 10% of children had very bad scores. Regarding physical QOL, 40% of children had good scores while 6,7% of them had very bad scores. Regarding emotional QOL 26.6% of children had a good score, 33.3% of them had a bad score. Regarding social QOL 30% had a very good score while 13.3 % had a very bad score. Regarding school QOL, 20%had a good score and 26.6% had a very bad score.

Age was significantly positively correlated with social and school aspects also with the total score of QOL **Table (6)**.

Table (1): Demographic data distribution among studied patients. (N=30)

		Mean	Median (Range)
Age		10.65±3.41	9.0(5-18)
		N=30	**100%**
Sex	• Female	10	33.3
	• Male	20	66.7

Table (2): Types of SCD among studied patients. (N=30)

Variant	N	%
SCA	20	66.7
SC trait	6	20
HbSβ^0 thalassemia	2	6.7
HbS/β^+ thalassemia	2	6.7

Table (3): Quality of life assessment distribution among studied group according to the questionnaire

		N	%
Physical aspect	**Good**	23	76.7
	Poor	7	23.3
Emotional aspect	**Good**	15	50.0
	Poor	15	50.0
Social aspect	**Good**	19	63.3
	Poor	11	36.7
School aspect	**Good**	14	46.7
	Poor	16	53.3
Total QOL	**Good**	19	63.3
	Poor	11	36.7

Table (4): Distribution of the physical activities, Emotional Functioning, Social Functioning and School Functioning of the studied patients

		Very good		Good		Fair		Bad		Very bad		Total
		N	%	N	%	N	%	N	%	N	%	
Physical Functioning	Walking	22	73.3	5	16.7	2	6.7	1	3.3	-	-	30
	Running	16	53.3	7	23.3	2	6.7	3	10	2	6.7	30
	Making activity	20	66.7	3	10	1	3.3	4	13.3	2	6.7	30
	Lifting something	15	50	4	13.3	2	6.7	2	3	10	4	30
	Taking a bath	25	83.3	3	10	1	3.3	-	-	1	3.3	30
	Doing chores	23	76.7	1	3.3	2	6.7	2	6.7	2	6.7	30
	No pain	13	43.3	5	16.7	7	23.3	3	10	2	13,3	30
	Having energy	8	26.6	7	23.3	8	26.6	3	10	4	13.3	30
Emotional Functioning	Being afraid	15	50	7	23.3	3	10	4	13.3	1	3.3	30
	Being sad	14	46.7	5	16.7	4	13.3	3	10	4	13.3	30
	Being angry	7	23.3	3	10	3	10	10	33.3	7	23.3	30
	Sleeping trouble	12	40	10	33.3	2	6.7	3	10	3	10	30
	Being worried	5	16.7	5	16.7	4	13.3	11	36.7	5	16.7	30
Social Functioning	Getting along with kids	21	70	6	20	2	6.7	1	3.3	-	-	30
	Other kids refuse him	28	93.3	1	3.3	1	3.3	-	-	-	-	30
	Teasing from other kids	28	93.3	2	6.7	-	-	-	-	-	-	30
	Can't do things as others	18	60	4	13.3	1	3.3	2	6.7	5	16.7	30
	Keeping up when play	20	66.7	5	16.7	2	6.7	2	6.7	1	3.3	30
School Functioning	Pay attention in class	7	23.3	4	13.3	3	10	11	36.7	5	16.7	30
	Forgetting things	-	-	4	13.3	6	20	12	40	8	26.7	30
	School work	6	20	3	10	2	6.7	2	6.7	17	56.7	30
	Missing school due to illness	7	23.3	2	6.7	3	10	3	10	15	50	30
	Missing school to go to the hospital	5	16.7	4	13.3	3	10	2	6.7	16	53.3	30

Table (5): The total QOL scale and QOL Subscales of the studied sickle cell patients

QOL	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	%	
Total QOL	6	20.0	9	30.0	4	13.3	8	26.6	3	10.0	36.4±13.21
Physical	7	23.3	12	40.0	4	13.3	5	16.7	2	6.7	10.73±3.12
Emotional	5	16.7	8	26.6	2	6.7	10	33.3	5	16.7	8.93±2.74
social	9	30.0	6	20.0	4	13.3	7	23.3	4	13.3	7.16±2.36
School	4	13.3	6	20.0	4	13.3	8	26.6	8	26.6	9.56±3.12

Table (6): Correlation between QOL scores and age, mother's age, and BMI

		Total	Physical state	Emotional state	Social state	School function
Age	**r**	0.444*	0.288	0.282	0.389*	0.542**
	P	0.014	0.123	0.131	0.034	0.002
	N	30	30	30	30	30
Mother age	**r**	0.264	0.187	0.177	0.304	0.234
	P	0.159	0.324	0.350	0.103	0.212
	N	30	30	30	30	30
BMI	**r**	0.357	0.345	0.202	0.279	0.352
	P	0.053	0.062	0.284	0.135	0.057
	N	30	30	30	30	30

(*) P value is statistically significant at <0.05 (**) P value is highly statistically significant at<0.01

DISCUSSION

The mean age of SCD patients in our study is 10.65±3.41years. This is in similarity with **Nafee and Salem** [6] in Mansoura University Hospitals who reported that the mean age was10.2 ± 0.9 years in their study.

Our study showed that males (66.7%). Our finding is supported by another study done in Macca region in Saudi Arabia that males are 67.5% and females are 32.5%. **Alharbi *et al.*** [7]**.**

Our study found that genotypes of sickle cell diseases are sickle cell anemia is 67%, 20% is sickle cell trait and 13% is sickle cell HbS/β^+ thalassemia. This finding of the present study goes in line with the finding of [8].

In our study, we used the WHOQOL-BREF criteria for measuring QOL in SCD and found that children with SCD scored positive 76.7% in physical activity, 50% in emotional aspect, 63.3%social aspect, 46.4% in school aspect, and the total QOL assessment is 63.3%.

Concerning **the physical aspect** of QOL in our study, our children had no problem with self-care followed by usual activities and mobility such as taking shower, doing chores around the house, and doing sports. These findings are not in agreement with the study of [8].

Concerning QOL for **emotional functioning**, the present study revealed that (56.7%) of the studied sample often worry about what will happen to him or her, (53.3%) feeling angry and feeling afraid or scared. These findings are in agreement with **Alharbi *et al.*** [7] who found that more than half of children with SCA always felt afraid or scared.

Our study showed that the majority of children 73.4% were sleeping well at night. This finding is nearly in agreement with **Barakat** [9] who illustrated that 56 % of children with SCA were sleeping well at night. On another hand, **Alharbi *et al.*** [7] found that more than half of children with SCA had always troubles in sleeping patterns.

The aspect of **social function** in our study revealed that 96.6% of sickled children never had a problem in kids' friendship and 90% sometimes got along with other children. This result disagrees with **Alharbi *et al.*** [7] where it cleared that the majority of the children with SCD always faced issues such as other children did not want to be their friends, having trouble getting along with kids. On the other hand, our result agrees with [6]**.**

Concerning **school achievements** of children with SCD, the results of the present study revealed that 53.3% of students didn't pay attention in class and 66.7% of the children forget things, which agreed with the results of **Alharbi *et al.*** [7]. Approximately, 70% of children had difficulty in keeping up with school work, and nearly about three quadrants of the studied group missed school to go to the hospital or doctor There were statistically significant differences in all school health subitems.

The results of the present study revealed that most children with SCD had either poor (36.7%) or good (63.3%) quality of life.

Sehlo and Kamfar [10] supported these findings by a study done in the King Abdulaziz University Hospital in Jeddah in Saudi Arabia where they mentioned that higher levels of parent support were significantly associated with decreased depressive symptoms, improve psychological aspect, and better quality of life in children with SCD.

Panepinto *et al.* [11] didn't support these findings where they mentioned that chronic health problems such as SCA are negatively affected the health-related QOL of children. Other authors also stated that most children cases with SCA during school years had low QOL.

CONCLUSION

It was concluded that sickle cell disease is a chronic disease that had a negative impact on QOL that includes physical, emotional, social, and school

functioning. Our results reflected that all aspects of QOL were affected especially school functioning.

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