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# Effect of Structured Sickle Cell Anemia Health Care Intervention Package on the Quality of Life of Sickled Children

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*Abstract:* Sickle cell disease (SCD) is an autosomal recessive disorder characterized by production of abnormal hemoglobin S, which associated with high morbidity and mortality. The highest prevalence of SCD in Saudi Arabia is in the Eastern province. SCD considered as a chronic disease which may affecting child's quality of life on all domains as physical, mental, school function and social well-being. Objectives: Assess quality of life of sickled children before and after implementation of structured sickle cell anemia health care intervention package. Subjects and Methods: a convenient sample of 30 children having sickle cell anemia their age ranged between 8-12 years were taken from out- patients clinic in AL-khobar City .Tools: An interview questionnaire sheet used to fulfill Child's socio demographic data and Pediatric Quality of life and WhatsApp application to follow up sickled children. Results: median of all dimensions of Quality of life score for sickled children (physical, emotional, social and school function) before implementing health care intervention package were 75.0 (6.3), 64.0 (10.0), 43.0 (10.0) and 72.5 (10.0) comparing to after implementation were 21.9 (6.2), 15.0 (10.0), 26.5 (5.0) and 30.0 (5.0) respectively and this improvement reflecting decrease number of hospitalization from 100% before implementation to 97% after. All sectors and agencies should be collaborating with these children and their families to improve their quality of life.

*Keywords:* Sickle cell anemia, Quality of life, intervention package, children.

# 1. INTRODUCTION

Sickle cell diseases (SCD) considered a group of genetic , autosomal recessive disorders that affects the hemoglobin There are three different types of SCD, sickle cell anemia (Hb SS), sickle hemoglobin C disease (Hb SC) and sickle beta thalassemia disease (Hb SbThalassemia).Centre of Children with Special Needs defined , sickle cell anemia (Hb SS) as a disease that "comprises a group of genetic disorders characterized by the inheritance of sickle hemoglobin (Hb S) from both parents and the child become affected where red blood cells distort into a sickle, or crescent, shape., or the child take it from one parent and being a trait carriers <sup>(1, 2)</sup> It affects individuals of African, Mediterranean, and Asian descent and considered as a chronic disease associating with high morbidity and mortality rates and adversely impact on the quality of life of sickle cell children <sup>(3)</sup>. In kingdom of Saudi Arabia the first cases of SCD was reported in eastern province in the 1960 s .The carrier status for SCD ranged from 2% to 27%, and up to 1.4% had SCD in Saudi Arabia and the prevalence in this province was highest comparing to others provinces and the main cause was consanguineous marriage which ranges from about 60% up to 90% in some Bedouin communities <sup>(4, 5, 6)</sup>

The clinical presentation of children with SCA varies greatly. The symptoms seen are usually the result of hemolysis of the cells and the compensatory mechanisms invoked by the subsequent anemia, and thrombi in the small vessels of various organs resulting from the sickling deformed sickle red cells which occlude the microvascular circulation producing vascular damage, organ infarcts, painful episodes and other symptoms associated with SCA. <sup>(7, 8)</sup>

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World Health Organization defined Health-related quality of life is commonly as the patient's physical, emotional and social well-being <sup>(9).</sup> The qualities of life of the children with Sickle Cell disease are largely affected due to recurrent severe pain crisis that shaping the daily life of children and continuous need for treatment, and care. The effects vary from economic, social, emotional, psychological, and activity issues. That means the child can't be able to practice his/her life normally as healthy age mate, whether in school or inside district area. <sup>(10,11)</sup>. The health-related quality of life (HRQoL) assessment provides a reasonable alternative to assess the impact of a disease process on the physical, social, and mental well-being of affected persons .It can also serve the purpose of evaluating the impact of the health interventions on SCA children.<sup>(12)</sup>

Interventions strategies to improve knowledge and skills of SCA children and their families that includes training in the recognition and management of SCA-related complications could provide patients abilities to cope with the disease symptoms, complications, and enhancing their families' confidence to determine early referral to healthcare providers that may improve clinical outcomes and appropriate decisions on whether referral is necessary and reduce the psychological reliance of the SCA children or their families on the healthcare professional<sup>.(13)</sup> So this study aims to study Effect of structured sickle cell disease health care intervention package on the quality of life of children with sickle cell disease.

### Aim of the study

Assess quality of life of sickled children before and after implementation of structured sickle cell anemia health care intervention package.

#### **Research hypothesis:**

- The sickle cell anemia affecting the quality of life of sickled children

- The quality of life of sickled children was improved after receiving a structured sickle cell anemia's health care intervention package.

## 2. SUBJECT AND METHODS

Research Design: A quazi experimental design was used to carry out the study.

Settings: The study was conducted in the out -patient clinic of King Fahd Hospital of University Al khobar City, kingdom of Saudi Arabia

**Sampling:** A convenient sample of 30 children having sickle cell anemia and their age ranged between 8-12 years were included.

**Exclusion Criteria:** children with other chronic diseases, handicapped and who had other forms of hemoglobinopathies were excluded.

#### Study tools:

Three tools were used for data collection:

**First tool** : An interview questionnaire sheet that developed by the researchers which included Child's socio demographic data as (gender and age, education) and data related to SCD as; when the child start to suffer from SCD, frequency of getting vaso-occlusive crisis.

**Second tool**: Pediatric Quality of life Inventory (Version 4)<sup>(14)</sup>, which adopted and included two parts:

- The first part: filled by the researcher through child's interview to assess their quality of life.
- The second part: filled by the child's parents/caregivers to report about their child's quality of life.

Four dimensions were included in Pediatric Quality of life Inventory (Version 4) for each parts; (physical, emotional, social and school functioning).

Each dimension also included several criteria for assessment of child quality of life with total number 23 criterion and presented in the form of Likert scale with five categories (1-5).<sup>(14)</sup>.

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- 4 Scoring system used in pediatric quality of life (PedsQL) Inventory with five categories (1-5) include:
- Category 1 takes 0 if the child never has a problem
- Category 2 takes 1 if the child sometimes has a problem
- Category 3 takes 2 if the child often has a problem
- Category 4 takes 2 if the child always has a problem
- Category 5 takes 2 if the child almost always has a problem

#### Total score =

**Third tool**: WhatsApp application which used for follow up child's health care plan and answering any questions from children / caregivers.

#### Methods:

### **Process of Study Implementation:**

- An official permissions were obtained from the responsible authorty in King Fahd Hospital of University (KFHU) ,Al-Khobar city for conducting the study.

### 1. Ethical considerations

- Ethical approval was asserted from University of Abdul Rahman Bin Faisal in KSA.
- Verbal approval was obtained from the parents/caregivers of children who were participated in the study.
- Confidentiality and anonymity of individual response was guaranteed
- Children and their parents were assured that the collected data used only for the purpose of research and they had the right to withdraw from the study at any time.

### 2. Literature Review:

A review of the international and national literature was carried out on the diverse aspects of SCD, and child quality of life, using scientific published articles, internet search, and textbooks.

### 3. Development of a structured health care intervention package for SCA:

Which developed by researchers and containing a designed booklet to cover all the needed knowledge, prevention, protection and health care education for children and their caregivers. Beside, initiating access for continues communication, counseling and follow up with children and their caregivers through process of social media such as WhatsApp.

### \*The developed booklet covered the following aspects:

- The disease process as predisposing factors ,pathophysiology ,signs / symptoms and complications
- Discharge plan of care, non-pharmacological management of sickle cell pain crises.

• Health education about importance of increasing fluid intake, proper feeding, use warm bath or warm compress, care of fever, skin redness, and swelling.

- Pharmacological management as use of antipyretic, analgesic and vitamins supplementation and use of hydroxyl urea.
- Counseling for infection prevention, travelling precautions, avoiding excretion /exposure to dehydration whatever the causes and blood transfusion, priapism, acute chest syndrome a being complication of SCA.

• Monitoring follow up care which covered by routine hospital follow up care schedule and follow up by weekly communication and contacting by the researchers using mobile phone and WhatsApp application for answering any SCA questions for caregiver /children.

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## \*Scoring system:

The PedsQL<sup>TM</sup> Generic Core Scale version 4.0 is a multi-dimensional child self-report and parent proxy report of generic core measures integrated with disease-specific modules. It has four domains: physical, emotional, social and school functioning, with child self-report versions for a broad age range (age groups 5–7, 8–12, and 13–18) and parallel parent proxy reports for each age group. Likert response scale items are reverse-scored and linearly transformed to a 0–100 scale (0 = 100, 1 = 75, 2 = 50, 3 = 25, 4 = 0), so that higher PedsQL<sup>TM</sup> scores indicates better HRQoL.

## 4. Study tools:

- Tools of data collection were adopted after reviewing the related literature except tool 1 which developed by the researchers. Validity was tested by 4 expertize in pediatric and community health nursing field.

**5. Pilot study:** A pilot study from 5 children with sickle cell disease was carried out in order to identify any needed modification and those children were excluded from the study sample.

### 6. Data collection:

- The purpose of the study explained to children and their caregivers. Through an interview questionnaire (tool 1) and pediatric quality of life inventory (PedsQLI) sheets (tool 2, part I); were used for data collection from children before and after implementation of a structured health care intervention package for SCA; the interview done by the researcher in the waiting area of the outpatient clinic and taken from 10-15 minutes for each interview.

- Data was collected from child's caregivers by using (tool 2, part II); whereas, they report upon their children quality of life before and after implementation of a structured health care intervention package for SCA. The researchers used small group discussion from 5 caregivers of SCA children in waiting area of outpatient clinic to discuss, clarify and cover the area of health education knowledge related to their children and it took from 20 to 35 minutes and the related information were written and illustrated in a booklet using a simple way and lifted to the caregivers.

### 7. Statistical Analysis

All categorical data were represented by frequency with percentage, and continuous data were presented by Median with inter quartile range because the percentage of each quality life is not follow the normal distribution. Demographic variables of SCA were compared by Chi-square test for categorical variables and Mann-Whitney U test for continuous variables. Also, Mann-Whitney U test was used for testing the significant difference of percentage of Quality life before and after implementation of health care intervention package. All the analysis was done by using SPSS 21.0 version A and p value less than 0.05 were considered as significant.

## 3. RESULTS

## Part I: Demographic data

Studied ample	Sickled children		
	(n =30)		
Variables			
Age			
Mean $\pm$ SD	$10.2\pm0.9$		
Median (IQR)	10.5(1.5)		
Min Max.	8.6 - 12		
Gender, n (%)			
Male	7(23.3)		
Female	23(76.7)		

## Table 1: Distribution of children according to their demographic data

Z = Mann-Whitney U test &  $\times^2 =$  Chi-square test

**Table 1:** illustrates the average age of sickled children was 10.2 with SD  $\pm$ 0.9 and 76.3% of them were females.

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Figure 1 and 2: Distribution of the number of hospitalization before and after implantation of SCD health care intervention package

## Figure 1:



Figure2:



**Figure 1 and 2 :** Shows frequency of hospitalization of the sickled children through previous three months before and after implementation of SCA intervention health care package, 47% compared to 23% of them were admitted more than 3 times; whereas be equal 30% more than two time and 23% compared to 40% of them were got once respectively.



Figure 3: Distribution of sickled children according to age of onset of disease

**Figure 3:** illustrates that the highest percentage was 63% of children suffering from the SCA at the age of 3 years where the lowest (17%) at the age of 4 years. In addition to 20% of them start suffering at early age of 2 years.

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## Part II: Pediatric Quality of life inventory (version 4).

## Table 2: Overall quality of life of sickled children before implementation of SCA health care intervention package.

1. Physical Functioning (PROBLEMS WITH)	Never 0	Almost Never 1	Some -times 2	Often 3	Almost Always 4
1.Walking more than one block	0(0.0%)	0(0.0%)	0.0	9(30%)	21(70%)
2.Running	0(0.0%)	0(0.0%)	3 (10%)	3(10%)	24(80%)
3.Participating in sports activity or exercise	0(0.0%)	0(0.0%)	0(0.0%)	0(0.0%)	30(100%)
4.Lifting something heavy	0(0.0%)	0(0.0%)	0(0.0%)	15(50%)	15(50%)
5.Taking a bath or shower by him or herself	30(100%)	0(0.0%)	0(0.0%)	0(0.0%)	0(0.0%)
6.Doing chores around the house	0(0.0%)	2(6.7%)	5(16.7%)	5(16.7%)	18(60%)
7.Having hurts or aches	0(0.0%)	0(0.0%)	8(26.7%)	22(73.3%)	0(0.0%)
8.Low energy level	0(0.0%)	0(0.0%)	13(43.3%)	9(30.0%)	8(26.7%)
2. Emotional Functioning (PROBLEMS WITH)	Never	Almost Never	Some -times	Often	Almost Always
1.Feeling afraid or scared	4(13.3%)	0(0.0%)	6(20.0%)	20(66.7%)	0(0.0%)
2.Feeling sad or blue	0(0.0%)	0(0.0%)	17(56.7%)	13(43.3%)	0(0.0%)
3.Feeling angry	0(0.0%)	0(0.0%)	9(30%)	21(70%)	0(0.0%)
4.Trouble sleeping	1(3.3)	0(0.0%)	9(30%)	20(66.7%)	0(0.0%)
5.Worrying about what will happen to him or her	0(0.0%)	0(0.0%)	3(10%)	27(90%)	0(0.0%)
3. Social Functioning (PROBLEMS WITH)	Never	Almost	Some	Often	Almost
		Never	-times		Always
1.Getting along with other children	7(23.3%)	0(0.0%)	12(40%)	11(36.7%)	0(0.0%)
2.Other kids not wanting to be his or her friend	30(100%)	0(0.0%)	0(0.0%)	0(0.0%)	0(0.0%)
3.Getting teased by other children	1(3.3%)	0(0.0%)	11(36.7%)	18(60%)	0(0.0%)
4.Not able to do things that other children his or her age can do	0(0.0%)	0(0.0%)	30(100%)	0(0.0%)	0(0.0%)
5.Keeping up with other children	0(0.0%)	0(0.0%)	30(100%)	0(0.0%)	0(0.0%)
4. School Functioning (PROBLEMS WITH)	Never	Almost Never	Some -times	Often	Almost Always
1.Paying attention in class	0(0.0%)	0(0.0%)	0(0.0%)	0(0.0%)	30(100)
2.Forgetting things	0(0.0%)	0(0.0%)	9(30%)	21(70%)	0(0.0%)
3.Keeping up with schoolwork	0(0.0%)	0(0.0%)	7(23.3%)	23(76.7%)	0(0.0%)
4.Missing school because of not feeling well	0(0.0%)	0(0.0%)	11(36.7%)	19(63.3%)	0(0.0%)
5.Missing school to go to the doctor or hospital	0(0.0%)	0(0.0%)	11(36.7%)	19(63.3%)	0(0.0%)

Statistically significant at 1% (highly significant)

**Table3:** shows overall quality of life of sickled children after SCA health care intervention package implementation ;regarding problem in physical function 100% of them never have a problem in bathing them self while the same percentage almost always had a problem sport activities .In the emotional aspect 90% of them often had warrying about what will happen to them but the dimension of social function ,all children 100 % often had a problem in their ability to do things that the same age groups can do it .the school function was affect where 1005 of the children almost always paying attention in the class while 63.3% often missing the school attendance due to going to doctor or hospital follow up or emergency visit .

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Table 3: Overall quality of life of sickled children after implementation of SCA health care intervention package.

1. Physical Functioning (PROBLEMS	Never	Almost	Some	Often	Almost
WITH)		Never	-times		Always
1. Walking more than one block	0(0.0%)	22(73.3%)	7(23.3%)	1(3.3%)	0(0.0%)
2.Running	0(0.0%)	23(76.7%)	-	7(23.3%)	0(0.0%)
3.Participating in sports activity or exercise	0(0.0%)	13(43.3%)	7(23.3%)	10(33.3%)	0(0.0%)
4.Lifting something heavy	0(0.0%)	2(6.7%)	0(0.0%)	28(93.3%)	0(0.0%)
5. Taking a bath or shower by him or herself	30(100%)	0(0.0%)	0(0.0%)	0(0.0%)	0(0.0%)
6.Doing chores around the house	1(3.3%)	24(80%)	5(16.7%)	0(0.0%)	0(0.0%)
7.Having hurts or aches	3(10%)	24(80%)	3(10%)	0(0.0%)	0(0.0%)
8.Low energy level	25(83.3%)	5(16.7%)	0(0.0%)	0(0.0%)	0(0.0%)
2. Emotional Functioning (PROBLEMS WITH)	Never	Almost Never	Some -times	Often	Almost Always
1 Feeling afraid or scared	0(0.0%)	6(20%)	24(80%)	0(0.0%)	0(0.0%)
2 Feeling sad or blue	10(66 7%)	9(30%)	11(36.7%)	0(0.0%)	0(0.0%)
3 Feeling angry	0(0.0%)	6(20%)	24(80%)	0(0.0%)	0(0.0%)
4. Trouble sleeping	5(16.7%)	17(56.7%)	8(26.7%)	0(0.0%)	0(0.0%)
5.Worrying about what will happen to him or her	0(0.0%)	24(80%)	6(20%)	0(0.0%)	0(0.0%)
3. Social Functioning (PROBLEMS WITH)	Never	Almost Never	Some -times	Often	Almost Always
1.Getting along with other children	0(0.0%)	24(80%)	6(20%)	0(0.0%)	0(0.0%)
2.Other kids not wanting to be his or her friend	0(0.0%)	26(86.7%)	4(13.3%)	0(0.0%)	0(0.0%)
3.Getting teased by other children	0(0.0%)	7(23.3%)	23(76.7%)	0(0.0%)	0(0.0%)
4.Not able to do things that other children his or her age can do	0(0.0%)	27(90%)	3(10%)	0(0.0%)	0(0.0%)
5.Keeping up with other children	0(0.0%)	26(86.7%)	4(13.3%)	0(0.0%)	0(0.0%)
4. School Functioning (PROBLEMS WITH)	Never	Almost Never	Some -times	Often	Almost Always
1.Paying attention in class	0(0.0%)	25(83.3%)	5(16.7%)	0(0.0%)	0(0.0%)
2.Forgetting things	23(76.7%)	7(23.3%)	0(0.0%)	0(0.0%)	0(0.0%)
3.Keeping up with schoolwork	9(30%)	17(56.7%)	4(13.3%)	0(0.0%)	0(0.0%)
4.Missing school because of not feeling well	0(0.0%)	30(100%)	0(0.0%)	0(0.0%)	0(0.0%)
5.Missing school to go to the doctor or hospital	0(0.0%)	23(76.7%)	7(23.3%)	0(0.0%)	0(0.0%)

**Table3**: illustrates overall quality of life of sickled children after implementation of SCD health care intervention package where the physical function were tilted by a problem in sport activities that categorized by almost never , some -times and often (43.4%,23.3% and 33,3%) respectively .Regarding their problem in emotional function 80% of them almost never Worrying about what will happen to him or but in social dimension ,90% of them almost never had a problem in their ability to do things that the same age groups can do it. The school function affected by absenteeism from school days by almost never and some -times (76.7% and 23.3%) respectively.

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 Table 4: Correlation of Quality of Life Inventory Scores (QLIS) among sickled children before and after implementation of SCD intervention health care package.

QLI score	QLI score before HCPI	QLI score after HCPI	Statistical Test	P value
Dimensions of Quality of life problems				
Physical functioning problems				
Mean $\pm$ SD	$74.3\pm4.9$	$20.6\pm3.4$	Z = 6.708	< 0.001**
Median (IQR)	75.0 (6.3)	21.9 (6.2)		
Min – Max	62.5 - 81.3	14.6 - 31.3		
Emotional Functioning problems				
Mean $\pm$ SD	$65.2\pm6.5$	$14.7\pm6.6$	Z = 6.703	< 0.001**
Median (IQR)	64.0 (10.0)	15.0 (10.0)		
Min – Max	50 - 75	0-25		
Social functioning problems				
Mean $\pm$ SD	$41.2\pm5.6$	$26.7\pm4.2$	Z = 6.529	< 0.001**
Median (IQR)	43.0 (10.0)	26.5 (5.0)		
Min – Max	30 - 50	20 - 35.0		
School functioning problems				
Mean $\pm$ SD	$73.7\pm4.9$	$26.3 \pm 4.9$	Z = 6.741	<0.001**
Median (IQR)	72.5 (10.0)	30.0 (5.0)		
Min – Max	65.0 - 80.0	20-35		
Total	65.7 (3.5)	22.8 (2.2)	Z = 6.677	< 0.001**

\*\*Statistically significant at 1% (highly significant), Z = Mann-Whitney U test

**Table 4:** illustrates that the overall problems in all dimensions of quality of life score were analyzed and compared before and after implementation of health care intervention package for sickled children where it was higher before implantation with 65.7(3.5); and 22.8(2.2) after implantation, with statistically significant relation where p <0.001.

\**Physical functioning:* The analysis also explored the median (IQR) quality of life scores related to problems in physical functioning for sickled children before health care intervention package was 75.0(6.3) after the implementation was 21.9(6.2) with highly statistically significant relation where p < 0.001.

\**Emotional functioning:* The median (IQR) quality of life score related to problems in emotional functioning for sickled children before health care intervention package was 64.0 (15.0) while after was 15.0 (5.0) with highly statistically significant relation where p <0.001.

\**Social functioning:* The median (IQR) quality of life score related to problems in social functioning for sickled children before health care intervention package was 43.0(10.0) compared to the after 26.5 (5.0) with highly statistically significant relation where p <0.001.

\*School functioning: This study also explored the median (IQR) score related to problems with school functioning for SCD children's before health care intervention package was 72.5 (10.0) while after was 30.0 (5.0) with highly statistically significant relation where p < 0.001

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 Table 5: Correlation between parents' report of their children's Quality of life inventory score before and after implementation of health care intervention package.

parents' QLIS report	QLIS before intervention package	QLIS after intervention Package	Statistical Test	P value
QL dimensions problems				
Physical functioning problems				
Mean $\pm$ SD	$73.7\pm4.9$	$27.3\pm4.8$	Z = 6.745	<0.001**
Median (IQR)	72.5 (10.0)	30.0 (5.0)		
Mini- Max	65-80	20-35		
Emotional functioning problems				
Mean $\pm$ SD	$79.5 \pm 3.6$	$21.8\pm3.1$	Z = 6.732	<0.001**
Median (IQR)	81.2 (3.9)	21.8 (3.9)		
Mini- Max	71 -84	16 -28		
Social functioning problems				<0.001**
Mean ± SD	$60 \pm 6.3$	$24.3\pm5.5$	Z = 6.751	
Median (IQR)	60.0 (6.2)	25.0 (6.2)		
Mini- Max	45-70	15-40		
School functioning problems				<0.001**
Mean $\pm$ SD	$79.5\pm6.7$	$27.2\pm5.0$	Z = 6.712	
Median (IQR)	80.0 (10.0)	27.5 (5.0)		
Mini- Max	65-90	15-35		
Overall				<0.001**
Mean $\pm$ SD	$73.4 \pm 2.5$	$24.0\pm2.9$	Z = 6.671	
Median (IQR)	73.9 (3.3)	23.9 (4.4)		
Mini- Max	68- 77	18-29		

\*\*Statistically significant at 1% (highly significant) & Z = Mann-Whitney U test.

**Table 5:** illustrates that median parents' report about overall problems in their sickled children in all dimensions of quality of life were analyzed and categorized in score before and after implementation of health care intervention package for SCD which was higher before 73.9 (3.3); than after 23.9(4.4), with statistically significant relation where p <0.001.

\**Physical functioning:* The parents median (IQR) quality of life score reported that the problems in physical functioning for their children before health care intervention package was 72.5 (10.0) while after was 30.0 (5.0) with highly statistically significant relation where p < 0.001.

\**Emotional functioning:* The parents median (IQR) quality of life score reported that their children problems in emotional functioning before health care intervention package was 81.2 (3.9) while after was 21.8 (3.9) with highly statistically significant relation where p < 0.001.

\**Social functioning:* The median (IQR) parents quality of life score report reflecting that their children had problems in social functioning before health care intervention package represented by 60.0 (6.2) compared to 25.0 (6.2) with statistically significant relation where p <0.001.

\**School functioning:* This study also explored the median (IQR) score parents ' quality of life score report about their children's problems with school functioning before health care intervention package was80.0 (10.0) while after was27.5 (5.0) with highly statistically significant relation where p < 0.001.

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# 4. **DISCUSSION**

The health-related quality of life (HRQoL) assessment provides a reasonable alternative to assess the impact of a disease process on the physical, social, and mental well-being of affected persons .It can also serve the purpose of evaluating the impact of the health interventions on SCD children Al Jaouni et al (2015)<sup>(12)</sup>. PQoLI Version 4 was used to determine the (QOL) of 30 children aged 8-12 years and their parents before and after implementation of SCA health care package.

The mean age was 10.2 years and the majority of sample was (76.7%) females. The quality of life of sickled children is deteriorated by episodic, debilitating pain which associated frequent hospitalization for disease complications. Regarding numbers hospitalization due to SCA complications; our study reviled that 100% from our studied sample complain from pain crisis and admitted to hospital before health care intervention package while after was minimized to be 97% and this result in concurrent with Karadağ etal (2018) <sup>(15)</sup> study that 98.5% from sickled children suffered from pain and admitted to hospital 1 to 5 times per year and decreased to be 95.1% after intervention health care program .

Our study found that SCD is associated with limitations in different aspects of HRQL, particularly physical, social, and emotional and school aspects. In all those aspects, PedsQL 4.0 scores were lower and this results in agreement with the findings of the following studies: Kipasika (2016) <sup>(16)</sup>, Sehlo & Kamfar (2016) <sup>(17)</sup>, and Menezes et al. (2014) <sup>(18)</sup> where health-related quality of life of children with SCD was poor all across the Pediatric Quality of Life Inventory score .The present study illustrated that the negative impact in the physical aspect of sickled children often was pain where (73.3%) of them complained from it as a complication that has the greatest impact on their HRQL and this result covenant with Al Jaouni etal (2013)<sup>(19)</sup> who explains the significantly lower HRQL score in patients who had pain crises.

On analysis of QOL for physical health subcategories for children with SCA, our study found that all sickled children (100%) almost always had a problem in sports activity or exercise, (80%) in running and (70%) Walking more than one block and these findings were supported by the study done by Elsayed and El-Gawad(2015)<sup>(20)</sup> Similar findings were observed in a study carried out by Asnani et al(2009)<sup>(21)</sup>. Concerning QOL for emotional functioning the present study revealed that (90%) of the studied sample often Worrying about what will happen to him or her, (70%) feeling angry and (66.7%)feeling afraid or scared and this findings in agreement with Alharbi1 et al (2016)<sup>(22)</sup> found that that 72.5% of children with SCA always felt afraid or scared. Similar results have been observed in the study done by Elsayed and El-Gawad (2015)<sup>(20)</sup>. The aspect of social function the present study found that (100%) of sickled children never had a problem in kids relation to be friends and (40%) some -times getting along with other children and this result disagree with Alharbi et al (2016)<sup>(22)</sup> 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.Concerning school achievements of children with SCA, the results of this study found that (100%) of them almost always was hardly paying attention in class, (76.7%) of them often had a problem in keeping up with Schoolwork and (63.3%) often missing school to go to the doctor or hospital and those findings correspondingly with Sadarangani et al (2018)<sup>(23)</sup> and Alharbi et al (2016)<sup>(22)</sup>

The professionals' responses mainly focused on preventive health through their description of temperature, hydration, and nutrition, safety with physical activities, taking folic acid supplements, and analgesia. In terms of the specific SCD-related problems, most of the professionals described similar strategies for pain management, namely, increasing oral fluid intake, applying heat, and topical medicines all of those are the recommended measures for healthy living and crisis prevention for SCD and so promoting a high quality of life. Forrester et al (2015)<sup>(24).</sup>

Crosby et al (2017)<sup>(25)</sup> and Molter & Abrahamson(2015)<sup>(26)</sup> emphasized that inadequate access to ambulatory care, poor communication with health care professionals, and lack of involvement in decisions regarding sickled children own health ,lacking of knowledge and comfort level of health care providers managing patients with SCD making a the gap in quality of life of those children .

Al Jaouni *et al*(2015)<sup>(19)</sup> who reported that treatment adherence and early intervention programs improves HRQoL of SCD children, Anie(2015)<sup>(27)</sup>also to hypothesis that educational interventions for the sickled children or caregiver may be beneficial in equipping them with the necessary skills to manage the physical and psychological demands of SCA ;and also Karadağ etal (2018)<sup>(15)</sup> reported that family education programs including parental courses, seminars, brochures, television programs and radio programs ,booklets and communication by mobile devices they can help children with SCA and their families deal with the undesired symptoms and disease-related complication the above three previous

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mentioned studies in agreement with our study results where all Median of dimensions of Quality of life (Physical ,mental ,social and school functioning) were improved after implementing a healthcare intervention package for SCA for sickled children and their families table (4).

Two trials reported on the parent's assessment of their child's QoL using the caregiver version of PedsQL in 44 children receiving intervention and 55 controls by Ketchen  $(2006)^{(28)}$  and Daniel $(2015)^{(29)}$ . Both trials included a face-to-face psychoeducational intervention ranging from one day (all four sessions lasting between 75 and 90 min done on a single day to reduce burden, followed by three 30-min booster phone call sessions) to six weeks. A fixed-effects meta-analysis showed that parents whose children received the intervention reported a lower score or improved QoL compared to the control group; however, this was not significant (no heterogeneity),MD -1.47 points (95%CI -5.41 to 2.47) Additionally, Daniel reported on child assessment of QoL (using the child version of the ' Peds QoLI and the reported improvement in QoL was 0.21 (P value = 0.60) and those trial in a congruence with our results where QLI score before implementing a healthcare intervention SCD package was 65.7 (3.5) and improved after the implementing a healthcare intervention SCD package to be 22.8 (2.2) table (4).

The present study illustrated that weekly monitoring of sickled children using WhatsApp application to send electronic message to patient's cell phone to

remind patient to take medication and answering any question may be the cause of improving QOL of them and the result in congruence with Badawy et al (2018)<sup>(30)</sup> recommended that a mounting evidence supporting use of mobile and technological access to improve and monitoring sickled children hydroxyurea adherence, QOL, and other important clinical outcomes.

Karadağ et al  $(2018)^{(15)}$  found that out of the children with SCA suffered from, 98.5% pain, 60.7% weakness and exhaustion, 20.9%, respiratory distress, 29.1% had partial difficulty in walking, 22.8% had partial difficulty in meeting their own needs and, to relieve their health problems, 72.3% of them could not go to school, 68.9% stayed in hospital at least 1-5 times a year while 48.5% used medications at home and all of these problems and needs in agreement with our study results before the implementation of the health care intervention package and the slightly improvement in QOL of after the implementation of the intervention denoted that in the light of these results, it is essential that family counseling centers for parents of children with SCA should be established, family education programs should be designed so as to ensure more attendance by families to these centers, and parents should be encouraged to communicate with other in addition to their adherence to comprehensive care by a multi-disciplinary team and preventive measures that minimize the mortality, morbidity and improve quality of life in sickled children .

# 5. CONCLUSION

As a result of this study (76.7%) of sickled children were females, most of the subcategories of the QOL of sickled children as physical, social, emotional, and school functioning were affected by SCA and median of QLIS in all QOL diminutions were improved after implementation of health care intervention package about SCA with statistical significant differences in addition to parents report QOLIS bout their sickled children all QOL diminutions were improved after the intervention sickle cell heath care package with statistical significant differences. Also hospitalization due to sickle cell pain crisis decreased from 100% before the health care intervention package implementation to be 97% after.

# 6. RECOMMENDATIONS

- [1] Developing an educational program for children with SCA /their parent through family centers care regarding risks factors, precipitating factors, clinical manifestations, prevention and management of sickle manifestations and complication.
- [2] Future research studies needed with larger samples of SCA and on other children age groups.
- [3] Further researches that collaborated by multidisciplinary team to study variables that affect QOL of sickled children.

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