Effect of instructional based nursing intervention program on self-efficacy, depression and quality of life in children with sickle cell disease

Nagat Farouk Abolwafa [(1)], Fatma Nagy Kotb [(2)], Howayda Mohammed Ali [(3)], Hala Samir Ahmed El-husseiny [(4)]

(1) Assistant professor of Pediatric Nursing, Faculty of Nursing, Minia Universities, Egypt
(2) Assistant professor of Psychiatric and Mental Health Nursing, Faculty of Nursing, Minia University, Egypt,
(3) Lecture of Pediatric Nursing, Faculty of Nursing, Minia Universities, Egypt
(4) Lecture of Pediatric Nursing, Faculty of Nursing, Damietta Universities, Egypt

Abstract

Background: The most prevalent inherited genetic illness in children is sickle cell disease (SCD). As they deal with the difficulty of recurrent painful exacerbations or crises, which can be life-threatening, children and adolescents with sickle cell disease may be at risk for psychological distress, including depression and low self-efficacy **(Benton, Boyd, Ifeagwu, Feldtmose, and Smith-Whitley, 2011). Objective:** is to evaluate the effect of instructional based nursing intervention program on self-efficacy, depression and quality of life in children with sickle cell disease. **Method:** The research design was a quasi-experimental (pretest as well as posttest) one. **Setting:** The investigation was carried out at the Minia University Hospital for Obstetrics and Pediatrics in pediatric hematology clinic. **Sample:** A purposive sample composed of 80 children with SCD from (10:18 years). **Tools:** fourth tools were utilized to collect data including, personal and clinical data sheet, Sickle cell disease self-efficacy (SCSES), Depression Self-Rating Scale for Children (DSRS-C), and World Health Organization Quality of Life-Brief (WHOQOL-BREF). **Results:** this study showed that there was positive improvement of children's level of self-efficacy and decreased depressive symptoms in post-test after 3months.also, children's knowledge and quality of life were enhanced in post/test after 3months with statistical significant difference between pre as well as post/test after 3months. **Conclusion:** The present study demonstrated that, instructional intervention program was effective in enhancing children self-efficacy which consequently decreasing depressive symptoms as well as better quality of life also the higher knowledge level. **Recommendations:** developing a continuous instructional program for interacting children's with sickle cell anemia in better self-management behaviors.

Keywords: depression, quality of life, self-efficacy, Sickle cell disease

Introduction

One of the common inherited blood problems is SCD. The shape of normal red blood cells are like the letter O and are soft, elastic discs that let them easily traverse a child's blood arteries. As contrasting with sickle cells are hard and viscous, and they are shaped same the letter C. Sickle cells are grouped jointly and adhere to the blood vessel lining, also it making challenging for them to pass via the narrow blood channels. The child's small blood capillaries are obstructed by these clusters, which slow down the flow of oxygen-rich, healthy blood. This restriction can reduce the amount of oxygen delivered to the tissues, that can lead to pain, dysfunction of the organ, also the most of diseases' sequelae **(Sundd, et al., 2019**). In addition to additional consequences including infections, stroke, acute discomfort on the chest, sluggish growth as well as children puberty, it primarily causes breathlessness, exhaustion, joint or organ pain, syndrome of the hand-foot, eyes issues, yellow eyes as well as skin, and other symptoms like yellow eyes and skin. Patients with SCD have a variety of distinct acute illnesses known as sickling crisis **(American Society of Hematology, 2019).**

Millions of people worldwide suffer from SCD, which has a significant impact on their morbidity and mortality. In the United States, there are around 100,000 people who have SCD **(National Center on Birth Defects and Developmental Disabilities, 2019)**. Globally, 300,000 infants are thought to be birth with SCD, with seventy-five percent of them occurring in Africa (**Piel, et al., 2013**). The care of indviduals with SCD may be split apart and drastically impacted by limited availability to healthcare team with the important competence in the SCD, despite pharmacological improvements aimed at disease modification and curative purpose**(Pai, et al., 2019).**

Self-efficacy(SE) is a key element in the management of chronic diseases because it mediates healthy behaviors and connect self-perceptions with person actions**(Frei, Svarin, Steurer-Stey &Puhan, 2009).** Additionally, the mechanism influencing the beneficial outcomes of self-management programs is SE. Promoting SE is crucial for developing self-management skills as a result **(Poku et al., 2018).** Moreover, understanding the warning signs and symptoms of a sickle cell pain crisis, being aware of potential side effects from SCD, and managing the condition on a daily basis are all examples of having SE in sickle cell patients**(Carey & Forsyth, 2019).** In a research by **Kanter et al. (2020),** patients with SCD reported more pain episodes than other participants, who had considerably higher SE.

One of the common prevalent mental well being conditions is depression(**Sundd, et al., 2019**). A few signs include anhedonia, sense of worthlessness, trouble focusing and sleeping, and suicidal thoughts. Research demonstrates that depression is a onerous circumstance with a poor effect on trajectories education, employment practice, as well as other life's areas. As stated by the World Health Organization(WHO), depression is a main reason of disability(**McHugh & Weiss, 2019**). In addition, A depressive illness was found in 4.6 percent of the 2,194 children as well as adolescents with SCD in a different retrospective investigation **(Jerrell, Tripathi, & McIntyre, 2011).**

There are measures of (QOL) quality of life available to evaluate the psychosocial effects of SCD. Due to recurrent pain crises that negatively impact their physical, mental, and psychosocial health, children with SCD typically have lower HRQOL than children of the same age who are in good health **(Beverung et al., 2014).**The same author said, "Studies evaluating children with SCD's QOL recommended that pain is the common significant predictor of a lower QOL among children with SCD." Additionally, compared to their healthy classmates, children with SCD showed worsening educational competency, according to several research**(Sarri, et al., 2018).** This also played a role in the perceived reduced QOL that SCD-affected youngsters reported. Additionally, SCD in the children had a bad mood had high acute discomfort, that had an adverse effect on their QOL.

The pediatric and psychiatric nurse's professional objective is to assist kids in engaging in a rank of therapies that are suitable for their clinical state. It is foreseeable that this technique will enhance health outcomes, reduce and stop unnecessary medical costs, and be implemented regularly and systematically. Mothers who participate in primary care and education have been shown to have children who are more developed cognitively, emotionally, socially, and physically; in public, interaction with health care teams is linked to a set of policies organizational that were estimated to ensure communication among these parties **(Murad, et al., 2019).**

Significant of the research:

According to research by **Pai et al. (2019),** SCD is characterized as a buildup or agglomeration of hereditary blood turbulence that is represented by the HbS hemoglobin variant. Unfortunately, up to 90% of these kids could pass away before turning 18 if they don't receive the right care. SCD alone accounts for 6.4% of under-five deaths in Tanzania **(Makani, 2020).** Additionally, **Suthan et al. (2020)** note that lack of knowledge about the condition has been associated with bad HRQOL results in children as well as adolescents with SCD, particularly given that knowledge of the condition is typically low among both afflicted patients and their careers.

Moreover, **(Ahmadi et al., 2015)** revealed that frequent referrals to the emergency room, together with unexpected and chronic pains, have caused sickle cell patients to have poorer self-esteem, a sense of frustration, sadness, worry, stress, and a poor QOL. Another study conducted by **Al-Azri et al. (2016)** revealed that the poor QOL experienced by children with SCD in Oman was mostly brought on by a lack of knowledge about the condition. In order to know infection signs and generally lifestyle adoption behaviors that avoid stirring up the illness, providing knowledge about chronic illnesses like SCD can improve and foster child as well as family health results. So, instructional programs can be meted through engagement as well as the education. By maintaining their independence, children can be assisted in having control over the welfare of their offspring **(Pai, et al., 2019**).

Therefore, developing and applying instructional program for children about SCD to improve SE and depression in children is prerequisite as well as advantages in terms related to the care quality to reduce morbidity and death rates, additionally to minimize families' responsibility, hospitals as well as the community.

The aim of the study:

The aim of this study is to evaluate the effect of instructional based nursing intervention program on SE, depression and QOL in children with SCD.

Research Hypothesis:

H1: Children with SCD who received the instructional based nursing intervention program will have a higher level of SE post program implementation.

H2: Children with SCD who received the instructional based nursing intervention program will have a lower level of depression post program implementation.

H3: Children with SCD who received the instructional based nursing intervention program will have a higher level of QOL post program implementation.

Subjects and Methods

Research design: The aim of this research was achieved through a pre as well as post-test "quasi-experimental" research methodology was utilized.

Operational definitions: For the sake of this research, the concepts listed below are defined based on prior research.

Sickle cell disease is the recurring episodes of discomfort and persistent anemia. The of the red blood cells shape is alternative from that of regular cells to that of a crescent or a sickle**(WHO, 2010).**

Sickle cell disease self-efficacy is a individual's confidence that, in spite of having SCD, they can carry out daily functioning tasks**(Jenerette & Murdaugh, 2008).**

Setting: This investigation was performed at the Minia University Hospital for Obstetrics and Pediatrics in pediatric hematology clinic for patients with SCD.

Sampling and population: a purposive sample made up of (80) SCD patients who were accepted into the pediatric hematology clinic using the following inclusive criteria: children aged ten to eighty years. **Exclusion criteria**; youngsters with sickle cell anemia mixed with another illness (as thalassemia), and a history of a past cerebrovascular injury. **To calculate a sample size**, 0.05 was chosen as the degree of significance level, 0.95 was chosen as the power, and an effect size of 0.25 was used in the power analysis. 80 SCD-affected children met the minimum sample size requirement.

The following sample calculation formula serves as the foundation for determining the sample size: http://www.ifad.org/gender/tools/hfs/ anthropometry).

$$N = \frac{t^2 \times p(1-p)}{m^2}$$

$$N = \frac{(1.96)^2 \times 0.061\,(1-0.061)}{0.05^2} \qquad N = 80$$

Description:

N= needed size of the sample

t= (standard level of 1.960) which the level of confidence at 95%

p= prevalence estimated of children in Minia University Hospital for Obstetrics and Pediatrics. 2022 (0.06)

m = margin of error at 5percent (value of standard 0.050)

Tools of research data collection:

Fourth tools were used in this study, Based on relevant literature, we gathered the following types of information for this research:

Tool I: It was divided into the following 2 parts:

Part I: Personal and clinical information: as children as well as age, also the sex, and the residence.

Part II: Children's knowledge assessment forms: All children took this portion as a pre- and post-educational test after three months. Children's understanding of sickle cell anemia as well as sickle cell

crisis, involving definition, causes, symptoms, risk factors, as well as medical treatment, and the health education to prevent sickle cell crisis, as well as children's understanding of measures taken to overcome sickle cell crisis, such as receiving immunizations, drinking a lot of liquids, getting required oxygen, etc.

Scoring system of children's responses were evaluated utilizing a model answer sheet that created by the researchers in accordance with the items on the interviewing questionnaire sheet. Correct answers take one score, while erroneous answers take zero score. The scores of the items were sum for each portion, and the total was classified by the number of things. Then calculated the mean as well as the standard deviation of these scores, along with a percent score. Less than 50percent of the overall children's knowledge was deemed unacceptable, whereas 50 percent and higher was deemed satisfactory.

Tool II: Sickle Cell Self-Efficacy Scale (SCSES) (Bandura, 1994). This scale, which consists of nine items, utilized a summated rating approach to assess a person's perception of their capacity to involve in everyday functioning exercise despite having SCD. Responses on the whole scale range from one equal ("not at all sure") to five equal ("very sure"); the measurement unit is the interval. A total score is calculated by adding the replies to each item; higher scores denote stronger SE. The total score was 45, the scores from (9-27)is lower SE) and scores from (28-45 is higher SE). The internal consistency reliability was .87.89 was documented lastly **(Edwards et al., 2000).**

Tool III: Depression Self-Rating Scale for Children (DSRS-C) (Birleson et al., 1978) which is a self-rating scale for children to measure the acuity symptoms of depression in children and adolescent aged from 8-16 years old. It consisted of eighteen items rated on a three point scale (zero point equal not true, one point equal sometimes true, as well as two point equal true). The total score on the DSRS ranges from 0-36 with higher scores indicating more sever symptoms of depression. With an alpha coefficient of 0.86, it has a good level of internal consistency, test reliability, as well as established validity. A psychiatric diagnosis of Major Depression was substantially more likely to be provided to kids with DSRS-C scores of 15 or higher. The Scale's test-retest reliability on a diverse sample revealed acceptable stability (0.80). The item scores were summed, as well as the amount was classified by the number of things, the scores from (0- 18 is lower depression) and scores from (19-36 is higher depression).

Tool IV: World Health Organization Quality of Life-Brief (1995) (WHOQOL-BREF) with Arabic version. There were twenty-six items, with twenty-four of them including 4 different QOL domains: physical health (7 items), as well as the psychological health (6 items), and the social interactions (3 items), additional (8 items) of the environment. Moreover 2 additional items evaluated overall QOL not included in the total score. In Questions 3, 4, and 26, three negatively phrased items were ranked on a 5-point response scale (from 0-5). Then calculated the mean of each domain score. High scores revealed high QOL. The scoring system from (0-100), the scores from (0- 50) is lower QOL) and scores from (51-100 is higher QOL).

Validity and Reliability:

Three specialists in the field served as the jury that examined the tools' content validity, and any necessary modification were made. Using a Cronbach's alpha coefficient, the tools' dependability was assessed to validate its consistency. The Cronbach's alpha for, WHOQOL-BREF, DSRS-C, SCSES (0.860, 0.860, 0.870); respectively coefficient approach was utilized to examine the consistency of the tools as well as reliability.

Pilot study of the research:

At Minia University Hospital for Obstetrics and Pediatrics, a pilot research with 8 children (10percent) was performed. A research pilot

study was carried out to assess the study tools' completeness and clarity and to ascertain the time needed to finish each tool. The essential modifications, omissions as well as additions were made in response to the pilot's results. Before beginning the actual study, the paperwork was authorized by the jury, and a pilot study was conducted to evaluate the reliability of the forms by calculating their internal consistency utilizing the a Cronbach's alpha coefficient.

Ethical consideration:

All children were given the opportunity to give their oral agreement to participate in the study after being informed of its aims. All voluntary participants were first introduced to the researchers, who gave them the assurance that the information they provided would remain completely private. They were made aware that participation in the study was completely voluntary as well as that they might revoke consent at any period of time. The privacy of the child's data was guaranteed. Anonymity as well as confidentiality were guaranteed.

Instructional program:

The goal of the instructional nursing intervention was to assess how it affected the SE, depression, and QOL of SCD in children. The program was implemented at Minia University Hospital for Obstetrics and Pediatrics' hematology clinic. It was completed during a six-month period, from December 2022 to May 2023; the program's execution took this amount of time. To enhance contact, 10 subgroups of SCD patients were created, each with 8 kids. For each patient category, the same program sessions were used. Each subgroup received a total of 8 sessions, each lasting somewhere from 35 to 45 minutes.

Instructional methods include group discussions, role acting, modeling, instructive images, movies, and giving connecting comments have been employed throughout the curriculum. The researchers employed several forms of reinforcement, including delivering various candies, toys, and emotional support in the form of encouraging comments. The researchers summarized the session's content and asked the kids if they had any questions before concluding each session. In addition, homework assignments for each session were given to the patients. A copy booklet of the program containing all the instructional materials is given to each participant. A recap of the lessons from the last sessions and the goals of the recent one normally comes at the beginning of each session. During program implementation, students who showed interest were motivated by being praised and/or recognized.

The program composed of 4 phases: **the assessment phase**; the pre-test was created then disseminated to gather the necessary information after the researcher provided them a full clarification of the research's background and goals. Anytime more information was required, the researcher was on hand to provide it. In order to evaluate the children's degree of SE, depression, and QOL, four tools were used. **The planning phase;** after assessing SE, depression, and QOL, the researcher created the program content and created an instructional booklet to help children with their Arabic booklet needs. Data about SCD based on an analysis of pertinent literature and online sources. The instruction program's material was developed depend on an evaluation of the students being examined and their actual educational needs.

The implementation phase

The process began with getting to know the kids at the Minia University Hospital for Obstetrics and Pediatrics' pediatric hematology clinic. After that, the topic material was organized through theoretical workshops.

The program sessions include the following:

Session 1: This session began with meeting of the studied children, clarifying the objective, duration, and time of the session. The researchers also, building rapport and relaxation atmosphere with children, explained the program purpose, sessions' place and content through direct personal communication.

Session 2: This session aimed at helping the patients to gain information about SCD as definition, causes, clinical manifestations, complications, management, as well as

health teaching for the children about how to control crisis of sickle cell.

Session 3: This session includes demonstration of self-management behaviors such as taking medication as prescribed, staying hydrate, and appropriate communication with health care provider toward any change, using positive reinforcement after achievement of goals. Provide patients with resources to support SE, as educational material, self-management tools and how access to support group

Session 4: This session includes, behavioral technique involving relaxation techniques, deep breathing, techniques of distraction such as mental imagery, as well as positive phrases repeating. Patients were asked to apply deep breathing as well as relaxation exercise twice a day.

Session 5: This session aimed at teaching children the importance of social support from others, active listening, showing empathy, how to seek emotional support from others, and provide resources of emotional support.

Session 6: This session includes demonstration of problem solving techniques through situations and role paying, and children were assisted to know obstacles to behavior alternative, and were acquainted to solve those barriers.

Session 7: this session aimed at revising the precious sessions and teaching children how to seek pleasure from certain activities.

Evaluation phase:

In this phase, the instructional nursing intervention program was done using the four tools for assessing the achievement of intervention objective three months later

Data analysis

The Statistical Package for Social Science (SPSS 20.0) was used to code, categorize, tabulate, and analyze the data that had been collected. Standard deviation and mean were used to express descriptive data. Frequency and percentage were used to express qualitative data. The connection between two qualitative variables was examined using the Chi square. The t test was used to compare quantitative continuous data while comparing the pre-post application's mean scores. To evaluate the associations between the quantitative variables, Spearman's rank correlation was used. In order to visualize the data, graphs were created in Microsoft Excel. A result that has a P-value of 0.05 is considered significant, while one that has a P-value of > 0.05 is considered non-significant.

Results

Table (1) shows that, 75% of children age from 16:18 years, with mean age (17.8 ± 0.6). 60% of children were male while 40% were female. Regarding residence, 75% of children were living in rural but 25% were living in urban.

Table (2) illustrates that, 62.5% of children were visited the hospital for crises from 1-5 times, and 75% of them had a Vaso-occlusive crisis

Figure (1) demonstrates that, consanguinity among parents of children have sickle cell disease, there were 87.5% Close consanguinity among parents.

Figure (2) illustrates that, there were 90% of children with SCD receive blood transfusion.

Table (3) reveals that 32.5% of children correct knowledge about meaning of sickle cell anemia in pre-test but 97.5% of children in post-test after 3months. Regarding children correct knowledge about signs as well as symptoms of SCD was 25% in pre-test but 97.5% in post-test after 3months. Moreover, 17.5% of children correct knowledge about treatment of SCD in pre-test and 95% in post- test after 3months. 5% of children correct knowledge about controlling of sickle cell crises in pre-test and 100% in post-test after 3months. The mothers of children with less than five children, however, showed a statistically significant difference in their knowledge of the meaning, symptoms, as well as treatment and the prevention of SCD (P. 0.05, 0.05,0.03and 0.03); respectively.

Table (4) indicates that, The total means scores of children QOL, and SE were improved in post-test more than pre-test while depression decreased in post-test after 3months than pre-test.

Table (5) highlighted that there were statistically significant differences between total

mean score of QOL, depression states, and SE level in post-test after 3months among studied children (p=<0.05, 0.05, & 0.03) respectively.

Table (6): demonstrates the statistical significance of the negative associations between children DSRS-C as well as the total mean score of the WHOQOL-BREF and SCSE in pre as well as post-test after three months (r=-0.426-** P=<0.000, r=-0.452- **P=<0.000); respectively. The strongest statistically significant link discovered between the total scores of children's WHOQOL-BREF as well as SCSES was present between the outcome of the pre-test as well as post-test after three months of the children's who have SCD (r=0.545**) P=0.000.

Table (1): Frequency distribution of sickle cell children according personal data (n= 80)

Items	No	%
Age in years of children:		
10: 15years	20	25.0
16:18 years	60	75.0
Mean ± SD of children age	17.8 ± 0.6	
Gender of children		
Male	48	60
Female	32	40
Residence		
Rural	60	75.0
Urban	20	25.0

*NS= Not statistically significant differences

Table (2): Frequency distribution of sickle cell children according clinical data (n= 80)

Items	No	%
Number of Hospital Visits for Crises (per year)		
0	8	10
1:5	50	62.5
6:10	22	27.5
Types of sickle cell crises		
Vaso-occlusive crisis	60	75
Splenic sequestration	12	15
Aplastic crises	5	6.25
Acute chest syndrome	3	3.75

*NS= Not statistically significant differences

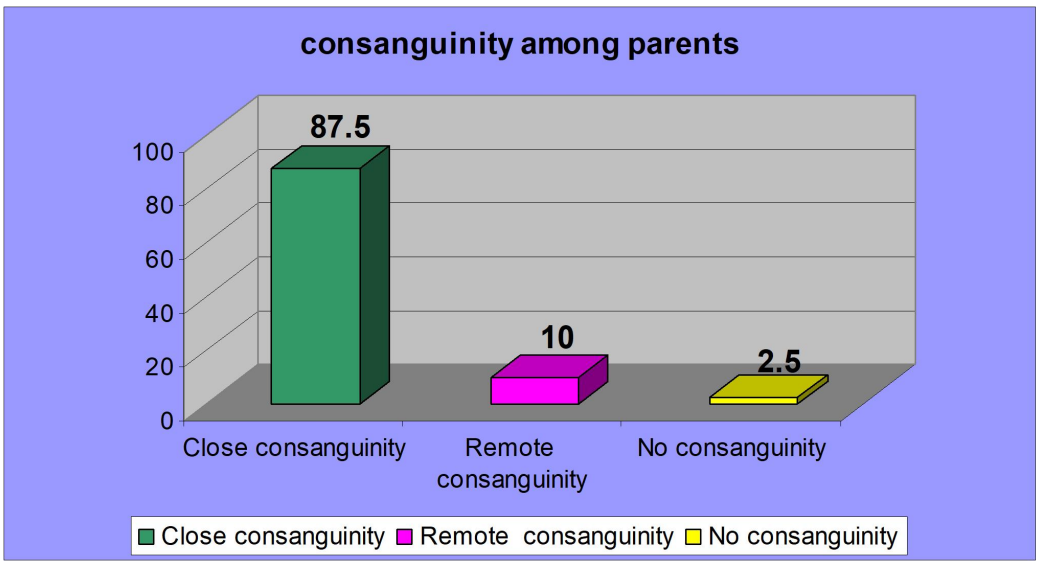

Figure (1): Frequency Distribution of consanguinity among parents

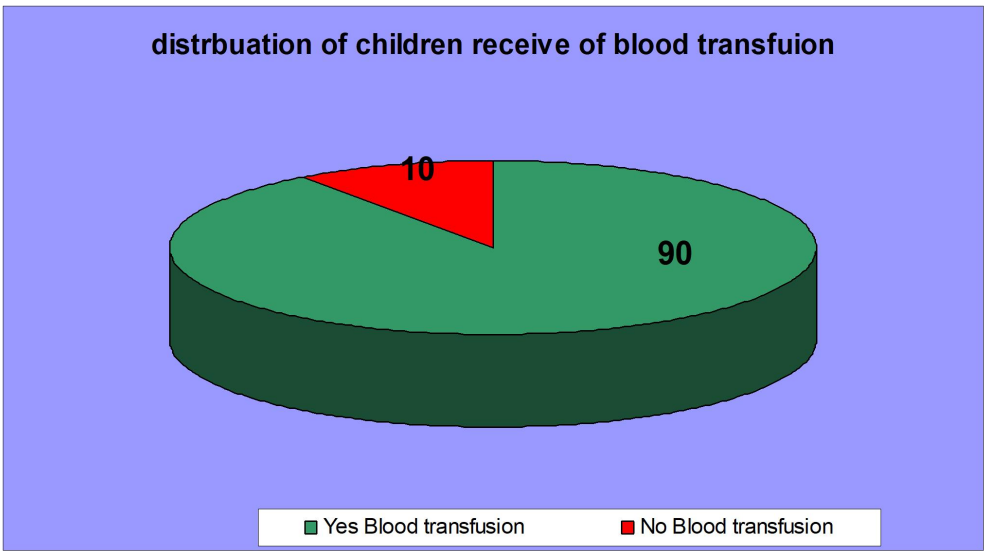

Figure (2): Frequency Distribution of children receive blood transfusion

Table (3): Distribution of children according to their knowledge about sickle cell disease in pre and post-test after 3months n=80

Items	Pre-test		Post-test after 3months		P. value
	No	%	No	%	
Meaning of sickle cell disease					
Correct	26	32.5	78	97.5	X2=0.62
Incorrect	54	67.5	2	2.5	0.05*
Causes of sickle cell disease					
Correct	13	16.25	70	87.5	X2=0.66
Incorrect	67	83.75	10	12.5	0.01*
Signs and symptoms sickle cell disease					X2=0.64
Correct	20	25	78	97.5	0.05*
Incorrect	60	75	2	2.5	
Treatment of sickle cell disease					
Correct	14	17.5	76	95	X2=0.68
Incorrect	66	82.5	4	5	0.03*
Preventions of sickle cell crises					
Correct	4	5	80	100	X2=0.60
Incorrect	76	95	0	0.0	0.03*
Management of Sickle cell crises					
Correct	10	12.5	80	100	X2=0.67
Incorrect	70	87.5	0	0.0	0.02*
Complications of Sickle cell disease					
Correct	15	18.75	72	90	X2=0.62
Incorrect	65	81.25	8	10	0.05*

*= Significant

Table (4): the total mean scores of higher (WHOQOL-BREF), lower (DSRS-C), and higher (SCSES) in pre and post-test after 3months (n=80)

	Pre-test	Post-test after 3months	t -test P. value
Variable	**Mean± SD**		
(WHOQOL-BREF	60.7 ± 13.3	85.28 ± 15.7	(0.03*)
DSRS-C	12.32 ± 12.2	10.32 ± 15.2	(0.05*)
SCSES	30.5 ± 15.3	44.36 ± 2.1	(0.05*)

Table (5): Total mean score of quality of Life, depression states, and self-efficacy level in pre and post-test after 3months among Studied Children (n=80)

	Pre-test	Post-test after 3months
Quality of Life		
Poor quality of life	26.32 ± 18.4	80.6 ± 13.2
high quality of life	60.7 ± 13.3	85.28 ± 15.7
t -test P. value	(0.03*)	(0.05*)
Depression states		
Lower depressive states	12.32 ± 12.2	10.32 ± 15.2
Higher depressive states	32.24 ± 2.4	11.45 ± 8.15
t -test P. value	(0.02*)	(0.05*)
Self-efficacy level		
Lower self-efficacy	20.36 ± 18.2	32.4 ± 12.8
Higher self-efficacy	30.5 ± 15.3	44.36 ± 2.1
t -test P. value	(0.04*)	(0.03*)

* Statistically significant at p=<0.05

Table (6): Correlation between mean scores of children's WHOQOL-BREF, DSRS-C, and SCSES in pre and post-test after 3months (n= 80)

Items		WHOQOL-BREF Pre-test	WHOQOL-BREF Post-test	(DSRS-C) pre-test	(DSRS-C) Post-test	SCSES Pre-test	SCSES Post-test
WHOQOL-BREF pre-test	R						
	P- Value						
WHOQOL-BREF post-test	R	**0.042****					
	P- Value	**0.000**					
(DSRS-C) pre-test	R	-0.631-	-0.305-				
	P- Value	.452	0.218				
(DSRS-C) Post-test	R	-0.245-	**-0.426-****	**-0.513-****			
	P- Value	.0.434	**0.000**	**0.001**			
SCSES Pre-test	R	0.343	**0.285****	-0.452-	-0.348-		
	P- Value	0.124	**0.000**	0.768	0.0823		
SCSES Post-test	R	0.456	**0.545****	-0.758-	**-0.452-****	**0.245****	
	P- Value	0.964	**0.000**	0.631	**0.000**	**0.001**	

Discussion:

Children with SCD suffer deficiencies in some areas of their health. Lower self-esteem, sadness, anxiety, and stress have developed in these cases as a result of unpredictable pain, frequent trips to the hospital as well as emergency rooms, and absence of employment (Ahmadi, et al., 2018). It goes without saying that such problems can affect SCD children's psychosocial functioning and lower their QOL in comparison to children without SCD (**Badawy, et al., 2021).**Numerous studies have shown that SCD children have low SE and poor QOL **(Bartlett et al., 2021).** The objective of this research is to assess how an instructional-based nursing intervention program affects children with sickle cell disease's SE, depression, and QOL

Regarding the studied children's data, the actually study findings proved that the majority

of children age from 16:18 years. These outcomes were unsupported by the research performed by **Anna et al., (2021)** who investigate biopsychosocial predictors of QOL in pediatric patients with SCD, the study results assured that 40percent of children's age ranked from 12.3 years, and their average age was (mean± SD = 12.3±2.1 years).

According to the results of the actually research, men made up the highest of participants in terms of gender. These findings were verified by a study performed by **Anna et al. in 2021,** which discovered that 60% of the children under observation were male. Despite the fact that this study was not comparable, **Yusra et al. (2021)** discovered that 68% of the youngsters were female and 32% were male.

Additionally, the results of the current study showed that more than fifty-percent of the children who were analyzed were from rural areas. This outcome may be explained by the fact that Minia University Hospital for Obstetrics and Pediatrics received nearly hospital referrals of acute cases for better facilities, technology, and care for children with different types of disorders in the Minia governorate and neighboring rural as well as areas of semi-urban. Also in the same context, 58% of respondents were from rural areas, while 42percent were from urban areas, according to **Nikita et al. (2022),** who discovered the locality distribution.

Moreover, the current study's findings showed that children visit hospitals on average 1:5 times per year for crises, with more than half of children having sickle cell crises. The preponderance of tight parental consanguinity occurred between parents of sickle cell disease patients. Approximately more than half of children with SCD suffer vaso-occlusive crises. Most youngsters who need blood transfusions do so. According to **Manase, et al. (2022),** a person with SCD experiences numerous vaso-occlusive episodes, necessitating frequent hospitalizations.

The present study finding proved that most of children have higher level of knowledge after instructional program intervention in post-test after 3months of intervention and engagement. This outcome is congruent with; **Shahine, *et al*., (2015)** who discovered that a reduced in the frequency of hospitalization between children with SCD was substantially associated with a rise in the caregiver's awareness and knowledge.

This study results evident that the total means scores of children SE were improved in post-test after 3months, which may be due to the positive effect of the program which help children to gain information about management of their illness, encouraged for self-management which help them to feel more control of their health. Also, helping them to set achievable goals that helping them to build confidence and feel a sense of accomplishment. This result is congruent with, **Mehrnaz et al., (2022)** who discovered SE or sense of coherence as predictors of health-regarding QOL in individuals with SCD, revealed that 19.3% of patients have low levels of SE. It is proposed to create specific health programs by concentrating on improving SE in SCD patients because this study indicated that increasing an individual's SE may increase their ability to take care of themselves. Another research by **Gomes et al., (2019)** who documented that, children who participate in the intervention program illustrated significant improvement in SE, health related QOL as well as psychological well-being. Also, this outcomes are consistent with **Gil et al (2009**) who investigate the impact of intervention program on SE in children with SCD, found that, children who participated in the program observed significant improvement in SE and self-esteem compared to those who did not shared.

As regarding depression, the actually research revealed that, that the total means scores of children depression was decreased after implementation of the program which may be related to the implementation of effective techniques as providing positive reinforcement to children with SCD when they successfully manage their symptoms which build confidence and reinforce positive behaviors. In addition, encouragement of social support, and connection with others who have the same condition, that, help children to feel less isolated and provide them with a support system, focusing on the positive aspect in their lives as achievement, hobbies, and addressing the emotional needs that related to their

condition through counseling, disclosure of emotion, relation exercise all of these behavior help to decreasing their depressive symptoms. This result is congruent with **Taiwo, et al., (2020),** who reported that, after intervention the mean score for depression was reduced from 22.13± 3.08 to 13.02± 4.56 which indicates significant improvement in depressive symptoms. In the same context, **Nikita et al., (2022)** The frequent vaso-occlusive crises, weariness, and lower working capacity that interfere with their ability to share in sports activities and school absences are the likely causes of the impact on the physical domain. Due to a variety of internalizing and externalizing behavioral issues, including anxiety, low self-esteem, and depression, the psychological development of children with SCD was impacted.

Furthermore, the present study shows that the total means scores of children QOL, were improved in post-test after 3months of instruional program more than pre-test. This result may be related to the impact of the instructional program through teaching children self-management, relaxation techniques, and emotional support, which help in improving their depressive symptoms pain crises which consequently improving their QOL. In the same context, **Taiwo, et al., (2020),** that documented that, after intervention the mean score for QOL was increased from 62.57 ±17.85 to 67.90±7.99 which indicates significant improvement in pediatrics QOL. Also, this result is consistent with (Murali et al., 2019), who reported the health related QOL score was significantly improved after the nurse led intervention among children with SCD.

The finding of current study illustrated that, there were statistically significant differences between total mean score of QOL, depression states, and SE level in post-test after 3months among studied children. Children who have instructional program have high level of SE in post-test after 3months. This result is congruent with **(Ahmadi et al., 2015)** that discovered that; the total score of QOL were significantly increased after intervention compared to those before intervention. In the same result **Al Nagshabandi and Abdulmutalib (2019)** who propose that specific health programs should be developed by concentrating on raising SE in SCD patients because increasing an individual's SE may increase their ability to care for themselves.

Conclusions:

The actual study illustrated that a higher level of children SE was significantly associated with reduced depressive symptoms as well as better QOL and higher level of knowledge after instructional program intervention in post-test after 3months of intervention and engagement.

Recommendations:

- Continuous developing instructional program for engaging children's with sickle cell disease in self-caring behaviors.
- Future studies on large sample are therefore, recommended for generalization of results about the effect of instructional nursing program on SE, depression, and QOL on children with SCD.

References

Abd El-Gawad, SME. (2017): Empowering Mothers to Overcome Sickle Cell Crisis in Their Children through Engagement and Education. *American Journal of Nursing Research, Vol. 5, No. 5, 182-190*

Ahmadi M, Poormansouri S, Beiranvand S, Sedighie L (2018): Predictors and correlates of fatigue in sickle cell disease patients. Int J Hematol Oncol Stem Cell Res 12:69–76

Al Nagshabandi EA, Abdulmutalib IAM (2019): Self-care management and self-efficacy among adult patients with sickle cell disease. Am J Nurs Res 7:51–57

Al-Azri MH, Al-Belushi R, Al-Mamari M, Davidson R, Mathew AC. (2016): Knowledge and health beliefs regarding sickle cell disease among Omanis in a primary healthcare setting: Cross-sectional study. Sultan Qaboos Univ Med J 2016; 16:e437 − 44. https://doi. org/10. 18295/ squmj. 2016.16.04.006.

American Society of Hematology (2019): CDC issues key clarification on guideline for prescribing opioids for chronic pain. https://www.hematology.org/ Newsroom /Press-Releases/2019/9537.aspx. Accessed 6 November 2019.

Anna M. H, Melanie K, Hanne S, Jamie K, April S, Baba I, Maria P, Jo H, Subarna C, Sue H, (2021): Biopsychosocial Predictors of Quality of Life in Paediatric Patients With Sickle Cell Disease. Frontiers in Psychology | www.frontiersin.org. September 2021 | Volume 12

Badawy SM, Beg U, Liem RI, Chaudhury S, Thompson AA (2021): A systematic review of quality of life in sickle cell disease and thalassemia after stem cell transplant or gene therapy. Blood adv 5:570–83. https:// doi. org/ 10. 1182/ blood advan ces.20200 02948

Bandura, A. (1994): Self-efficacy. In V. S. Ramachaudran (Ed.), *Encyclopedia of human behavior* (Vol. 4, pp. 71-81). New York: Academic Press. https:// www. phenxtoolkit. org/protocols/view/820401

Bandura, A. (1994): Self-efficacy. In V. S. Ramachaudran (Ed.), *Encyclopedia of human behavior* (Vol. 4, pp. 71-81). New York: Academic Press.

Bartlett R, Ramsay Z, Ali A, Grant J, Rankine-Mullings A, Gordon- Strachan G, Asnani M (2021): Health-related quality of life and neuropathic pain in sickle cell disease in Jamaica. Disabil Health J 14:101107. https:// doi. org/ 10. 1016/j. dhjo. 2021. 101107

Benton, T.D., Boyd, R., Ifeagwu, J,. Feldtmose, E., & Smith-Whitley, K. (2011): Psychiatric diagnosis in adolescents with sickle cell disease: A preliminary report. Current Psychiatric Reports, 13, 111-115. doi:10.1007/s119 20-011-0177-3

Best Bets, (2010): How long should an average sickle cell crisis last?" Archived from the original on 2010-06-17. Retrieved 2010-11-27.

Birleson P. Hudson I, Grey-Buchanan D, Wolff S. (1978): Clinical Evaluation of a Self-Rating Scale for Depressive Disorder in Childhood (Depression Self-Rating Scale). J. Child Psychology Psychiatry 28, 43/60.

Edwards R., Telfair, J., Cecil, H., & Lenoci, J., (2000): Reliability and validity of a self-efficacy instrument specific to sickle cell disease. *Behaviour Research and Therapy, 38*, 951-963.

Frei, A., Svarin, A., Steurer-Stey, C. & Puhan, M., (2009): Self-efficacy instruments for patients with chronic diseases suffer from methodological limitations – a systematic review. *Health and Quality of Life Outcomes*, *7*, 1-10. doi:10.1186/1477-7525-7-86

Gil K., Wilson J., Edens J., & Malcarne V., (2009). Effect of psychoeducational intervention on the adjustment and self-efficacy of children with SCD and their parents, Journal of pediatric psychology, 34(6), 677-688

Global Health Data Exchange. GBD Results Tool., (2019): Available online: http:// ghdx.healthdata. org/gbd-results-tool? params = gbd-api-2019-permalink/ d78 0 d f f be8a381b25e1416884959e88b (accessed on 8 August 2020).

Gomes M., Souza D., & Pinto, M., (2019): Cognitive behavioral intervention on quality of life and self-efficacy in children with SCD. European Journal of pediatrics, 178(5), 695-702.

Jerrell, J.M., Tripathi, A., & McIntyre, R.S (2011): Prevalence and treatment of depression in children and adolescents with sickle cell disease: A retrospective cohort study. Primary Care Companion CNS Disorders, 13(2), 93-97. doi:10. 4088/ PCC. 10m01063

Shahine R, Badr LK, Karam D, Abboud M. (2015): Educational intervention to improve the health outcomes of children with sickle cell disease. J Pediatr Health Care 2015; 29:54 – 60. https:// doi. org/ 10. 1016/ j. pedhc. 2014. 06.007.

Kendall, K.M.; Van Assche, E.; Andlauer, T.F.M.; Choi, K.W.; Luykx, J.J.; Schulte, E.C.; Lu, Y. (2021): The genetic basis of major depression. Psychol. Med. 2021, 51, 2217–2230. [CrossRef]

Kumar, Vinay; Abbas, Abul K., (2009): Fausto, Nelson; Aster, Jon Robbins and Cotran Pathologic Basis of Disease, Professional Edition: Expert Consult – Online (Robbins Pathology) (Kindle Locations 33498-33499). Elsevier Health. Kindle Edition.

Makani, J. (2020): Curative options for sickle cell disease in Africa: Approach in Tanzania. Hematol. Stem Cell Ther., 13, 66–70.[CrossRef] [PubMed]

Manase K. , Dorkasi L. Mwakawanga, Fatuma F. F. , Hamu J. Mlyuka, Lulu C. et al., (2022): The Effects of Sickle Cell Disease on the Quality of Life: A Focus on the Untold Experiences of Parents in Tanzania. Int. J. Environ. Res. Public Health 2022, 19, 6871. https:// doi. org/ 10. 3390/ ijerph19116871 https:// www. mdpi. com/ journal/ ijerph

Maurer M, Dardess P, Carman, KL., (2012): Guide to Patient and Family Engagement: Environmental Scan Report. (Prepared by American Institutes for Research under contract HHSA 290-200-600019). AHRQ Publication No. 12-0042-EF. Rockville, MD: Agency for Healthcare Research and Quality.[9]

McHugh, K.R.; Weiss, R.D. (2019): Alcohol Use Disorder and Depressive Disorders. Alcohol Res. 40, arcr.v40.1.01. [CrossRef]

Mehrnaz A, Samira B, Saeed P, Mahsa M, Camelia R, (2022): Sense of coherence or self‐efficacy as predictors of health‐related quality of life in sickle cell disease patients Annals of Hematology: https:// doi. org / 10. 1007/ s00277-022-05007-2. Received: 11 August 2022 / Accepted: 10 October 2022

Moez P., & Younan DN., (2016): High prevalence of haemoglobin S in the closed Egyptian community of SIWA Oasis. J Clin Pathol. 69(7): 632-6. Doi: 10. 1136/ jclinpath- 2015-203199.

Murali K., Arulppan J., & Thomas D., (2019): Effectiveness of led intervention on health related quality of life among children with sickle cell disease in Oman.

Myrvik, M.P., Campbell, A.D., Davis, M.M ,. &Butcher, J.L. (2011): Impact of psychiatric diagnosis on hospital length of stay in children with sickle cell anemia. Pediatric Blood and Cancer, 57, 297-302. doi:10.1002/pbc

Murad MH, Liem RI, Lang ES,. (2019): sickle cell disease guidelines by the American Society of Hematology: methodology, challenges, and innovations. Blood Adv.;3(23):3945-3950

National Center on Birth Defects and Developmental Disabilities. (2019): Data & statistics on sickle cell disease. https://www.cdc.gov/ ncbddd/sicklecell/ data. html. Accessed 3 October 2019.

Nikita S G, Chandrakant M. Bokade, (2022): Health related quality of life in children and adolescents with sickle cell disease in central India. International Journal of Contemporary Pediatrics *Gupta NS et al. Int J Contemp Pediatr. 2022 Mar; 9(3):268-273* http://www.ijpediatrics.com

Piel, F.B.; Patil, A.P.; Howes, R.E.; Nyangiri, O.A.; Gething, P.W.; Dewi, M.; Temperley, W.H.; Williams, T.N.; Weatherall, D.J.; DPhil, S.I.H.(2013): Global epidemiology of Sickle haemoglobin in neonates: A contemporary geostatistical model-based map and population estimates. Lancet, 381, 142–151. [CrossRef]

Poku, B., Caress, P., & Kirk, S., (2018): Adolescents' experiences of living with sickle cell disease: An integrative narrative review of the literature. International Journal of Nursing Studies, Vol.80 (No1), Pp: 20-28, https:// doi. org/ 10. 1016 / j. ijnurstu. 2017.12.008.

Sahoo, S.; Malhotra, S. (2018): Antecedents of depression in children and adolescents. Ind. Psychiatry J., 27, 11–16. [CrossRef]

Sarri G, Bhor M, Abogunrin S, Farmer C, Nandal S, Halloway R. (2018): Systematic literature review and assessment of patient-reported outcome instruments in sickle cell disease. Health Qual Life Outcomes. 2018;16(1): 99. PubMed| Google Scholar

Suthan P., Kamala M., Judie A. Sulaiman D. A. (2020): Health-Related Quality of Life of Children and Adolescents with Sickle Cell Disease in the Middle East and North Africa Region. Sultan Qaboos University Med J, November 2020, Vol. 20, Iss. 4, pp. e280–289, Epub. 21 Dec 20. https:// doi.org/ 10.18295/squmj.2020.20.04.002

Sundd, P.; Gladwin, M.T.; Novelli, E.M. (2019): Pathophysiology of Sickle Cell Disease. Annu Rev. Pathol., 176, 139–148. [CrossRef] [PubMed]

Taiwo T., Atilola O., Ani C., & Bola A., (2020): Effect of behavioral therapy on depression in adolescent with sickle cell disease, Open Journal of Psychiatry, Vol.10, No3 July2020.

World Health Organization Quality of Life-Brief (WHOQOL-BREF) (1995): position paper from the World Health Organization. Soc Sci Med. 1995 Nov; 41(10):1403–9. https://doi.org/10.1016/0277-9536(95)00112-K PMID:8560308

Yusra A, Eufemia J, Eunice L, Adey N, Mary-Lynn B, Wendie A and Adhra A. (2020): Parent educational intervention program for improving parental knowledge, self-efficacy and health related quality of life in children with sickle cell disease using smartphone technology: A randomized controlled trial. *Hematol Med Oncol,* 2020 doi: 10.15761/HMO.1000203. Volume 5: 1-8