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Quality of Life Based Education Program for Mothers of Children Suffering from Congenital Adrenal Hyperplasia

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Abstract: **Congenital adrenal hyperplasia (CAH) is a rare disorder characterized by cortisol deficiency with/ without aldosterone deficiency, and androgen excess. The aim of this study was to evaluate the effect of the educational program on quality of life for mothers of children suffering from congenital adrenal hyperplasia. A quasi-experimental design was used. This study was carried out at Benha University Hospital, Benha Specialized Hospital for Children and Health Insurance Hospital affiliated to Ministry of Health. A convenient sample included all available mother`s (55) of children between the ages of birth and 16 years with congenital adrenal hyperplasia. Tools of data collection: Three tools were used; Tool I: A structured interviewing questionnaire, Tool II: Mothers' reported practice sheet, Tool III: Quality of Life Scale. Results was a highly statistically significant difference observed between the studied mothers total knowledge, quality of life and mothers reported practice regarding to congenital adrenal hyperplasia at pre and post of the educational program implementation P= (<0.001). This study concluded that: The mothers of children with congenital adrenal hyperplasia had improved in their quality of life as well as the mother`s had a good knowledge and practice mothers of children suffering from congenital adrenal hyperplasia. Recommendations: The study recommended that future studies should emphasis on mothers` experience and management would enhance the current state of the science and provide a much-needed window into interventions aimed to improving the life of families and children with CAH.**

Keywords: **quality of Life, congenital adrenal hyperplasia, mothers reported practice.**

1. INTRODUCTION

Congenital adrenal hyperplasia (CAH) is a rare disorder characterized by cortisol deficiency, with/ without aldosterone deficiency, and androgen excess. Its a rare, inherited, genetic disorder that affects approximately 1 in 15,000 live births in the classical (salt-wasting) form, which is the most severe form of the disorder. The most common cause of CAH is 21-hydroxylase enzyme deficiency, which is characterized by cortisol deficiency, with or without aldosterone deficiency, and androgen excess.(**Louise et al 2011)**

Children with congenital adrenal hyperplasia (CAH) are at a significant risk for adrenal crisis, due to glucocorticoid and mineralocorticoid deficiency, during times of acute illness such as fever, vomiting, and accidental trauma, which are conditions commonly experienced by the general pediatric population. Adrenal crisis is an abrupt, life-threatening state that is caused by insufficient production of cortisol. Symptoms include hypotension, weakness, headache, vomiting, fever, chills, tachycardia, and shock. (**Beltz et al 2011)**

Management of CAH requires parents to administer oral steroids, typically hydrocortisone, up to three times daily. If a child is prescribed too much hydrocortisone, side effects can include growth suppression, obesity. If the dose of hydrocortisone is not sufficient, children with CAH are at a high risk for precocious puberty, which can also lead to stunted growth. Determining the proper dosage of steroids is typically achieved by obtaining routine laboratory work and measuring height, weight, and bone age every 3–6 months in a growing child**.(Boyse et al 2014)**

Additionally, stress dosing (doubling or tripling the oral steroid dose) is required when the child is experiencing an acute illness, such as fever or a broken bone. An emergency intramuscular (IM) injection of hydrocortisone is required when a child is unable to tolerate oral medications and/or if signs of adrenal crisis are present. The need for stress dosing, either orally or by injection, related to simple viral and bacterial childhood illnesses is frequent and unpredictable, often requiring parents to make complex treatment decisions**.(Gilban et al 2014**).

Mothers of children with CAH reported misconceptions and confusion about medication administration and general knowledge of the disorder. The mothers should observe the child closely for signs and symptoms of adrenal crisis, stress dosing when child is febrile also report signs and symptoms of reaction immediately to physician to minimize consequences, acute reaction that may occur at any time during the injection hydrocortisone **(Idris et. al 2014)**

Significant of study:

Congenital adrenal hyperplasia (CAH) is a rare, life threatening, endocrine disorder that affects boys and girls equally (**Louise et al 2017)**. Congenital adrenal hyperplasia have a negative impact on the quality of life for children due to the effects of the disease and its treatment, not only affecting children physical function, but also their social, emotional. This educational intervention was intended to improve quality of life for children suffering from CAH, which may help them live a better life and achieve a good QOL for them. The children with congenital adrenal hyperplasia were at high risk for many complications due to the lack of knowledge and inadequate care given to these children hence, the present study is under taken to evaluate the effect of the educational program on quality of life for mothers of children suffering from congenital adrenal hyperplasia

Aim of the study:

The aim of this study is to evaluate the effect of the educational program on quality of life for mothers of children suffering from congenital adrenal hyperplasia

Research Hypothesis:

Mothers' quality of life for children suffering from congenital adrenal hyperplasia significantly will be improved after implementing of the educational program.

Subjects and method:

Research Design:

A quasi- experimental research design was utilized.

Settings:

This study was carried out at the outpatient and Inpatient pediatric Endocrinology unites in Benha University Hospital , Benha Specialized Hospital for Children and Health Insurance Hospital affiliated to Ministry of Health .

Subjects:

A convenient sample of 55 mothers of children between the age of birth and 16 years with congenital adrenal hyperplasia disease.

Benha University Hospital(15) cases , Health Insurance Hospital(20) cases and (20) cases Specialized Pediatric Hospital at Benha City.

Tools of Data Collection:

In order to fulfill the aim of the present study, three tools were used for data collection:

Tool I : A structured interviewing questionnaire: It was developed by the researchers after reviewing related literature. It was written in Arabic language and composed of two parts to assess the following:

Part (1) : Personal characteristics of the studied sample which include:

a) Mothers' characteristics such as age, level of education, and residence.

b) Children' characteristics such as age, and gender, educational level and Child's rank.

c) Family and illness history: consanguinity relation between the father and the mother, presence of family members' relatives suffering from congenital adrenal hyperplasia and number of injectable hydrocortisone

Part (2): It concerned with mothers' knowledge regarding congenital adrenal hyperplasia, the true/false questions related to general knowledge of CAH (definition of congenital adrenal hyperplasia, definition of adrenal Crisis, causes of CAH, Signs and symptoms of CAH, treatment of congenital adrenal hperplasia, rout of administration hydrocortisone, side effect of over dose hydrocortisone and side effect of dose hydrocortisone is not sufficient

Scoring system for knowledge: The mothers' answers were compared with the model key answer, where 2 scores was given for complete correct answer, 1 score was given for incomplete correct answer and 0 score for wrong answer and unknown answer. The total score of studied mothers' answers were (8questions) to be 16 scores. The total score for all knowledge was classified as the following:

- Good knowledge ≥ 75%

- Average knowledge <50-75%

- Poor knowledge <50%

Tool II : Mothers' reported practice sheet.

It will be adopted from **Wilson(2008) & Hockenberry et al (2005)** to assess mothers` reported practice towards care of children with congenital adrenal hyperplasia ,as administer oral steroids, measuring pulse rate, measuring weight, measuring height and measuring fever. The total practices were 5 practices. It included the following practices: which include (26) items as the following:

1- Administer oral steroids (4 items)

2- Measuring pulse rate (6 items)

3- Measuring weight (4 items)

4- Measuring height (6 items)

5- Measuring fever (6 items).

Scoring system of the studied mother's reported practices was calculated as correct practice was scored 2 while correct and incomplete was scored 1 and wrong or incorrect was scored 0. The total score for all practice was classified as the following:

- Good reported practice ≥ 75%

- Average reported practice<50-75%

- Poor reported practice <50%.

Tool III : Quality of Life Scale:

This scale was adopted from ***de Jong et al (1997)***. to assess the quality of life for children suffering from congenital adrenal hyperplasia. It includes physical Wellbeing, psychological & mental wellbeing domains, social wellbeing domains, medication effect domains .The scoring system for each sub item was scored as, if it is never a problem (0), if it is almost never a problem (1), if it is sometimes a problem (2), if it is often a problem (3) and if it is almost always a problem (4) and categorized into either:

Good = 75- ≤100%

Average = 50- <75%

Poor = 0- <50%

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Preparatory phase:

Validity and Reliability

The researchers reviewed the past, current regional and international related literatures covering all aspects of the study using textbooks, articles, journal and scientific magazines. This helped the researchers to be acquainted with the research problem and guided them in developing the study tools. To measure content validity of the study tools, the researchers assure that items of the tools were adequately represent what are supposed to measure by presented it to three experts including; two in Pediatric nursing from the Faculty of Nursing Benha University, and one in Endocrinology from the Faculty of medicine Benha University, to test the content validity. Modifications of the tools were done according to the experts' judgment on clarity of sentences, appropriateness of contents and sequence of items. The experts' agreed on the content, but recommended minor language changes that would make the information more clear and precise. The suggested changes were made. Internal consistency reliability of all items of the tools was assessed using Chronbach's Alpha test. It was 0.83 for the structured interviewing schedule, and 0.86 for nurses' observation checklist.

2. METHOD

Exploratory phase:

Ethical considerations and human rights:

An official permission to conduct the study was obtained from the hospital mangers. Then participation in the study was voluntary; each mother was informed about the purpose, procedure, benefits, and nature of the study and each mother had the right to withdraw from the study at any time without any rationale, then oral/written consent obtained from them. Subjects were informed that obtained data will not be included in any further researches. Confidentiality and anonymity of each subject was assured through coding of all data and all information has taken was protected.

Pilot Study:

It was conducted on 10 % of the total study sample (5 mother`s) to evaluate the feasibility, reliability, and clarity of the tools .It was conducted to test the applicability of the tools, find out the possible obstacles and problems that might face the researchers and interfere with data collection. Additionally, detect any problems peculiar to the statements as sequence of questions and clarity. It was also helped to estimate the time needed for data collection, as it was 20 minutes.

The Educational Program Intervention

The aim of this study is to evaluate the effect of the educational program on quality of life for mothers of children suffering from congenital adrenal hyperplasia.

Conduction of theoretical part was performed through lectures and group discussions which were done in 3 sessions, (each session lasted for 15-20 minutes) and covered the following items. The sessions covered the items of the instruction guideline (definition of congenital adrenal hyperplasia, definition of adrenal crisis, causes of CAH, signs and symptoms of CAH, treatment of congenital adrenal hyperplasia, rout of administration hydrocortisone, side effect of over dose hydrocortisone and side effect of dose hydrocortisone is not sufficient)

Then conduction of practical part began at the previously mentioned settings. It was taken in 4sessions (each session for one hour) and covers the items.

First session: the primary assessment.

Second session*:* (administer oral steroids and measuring pulse rate,).

Third session: (measuring weight and measuring height)

Fourth session: (measuring fever) and secondary assessment

The teaching methods used were brainstorming lectures, group discussion, demonstration and re demonstration, role play and handouts. The program intervention carried out in the following four phases:

Field of Work:

Data were collected from the beginning of January 2018 to the end of December 2018. Immediately after the ethical approval was obtained; the researchers obtained oral consents from mothers who included in the study after an explanation of the aim, tools, benefits and the duration of the study to gain their cooperation. The researchers then started to interview each mothers individually and this took about 15-20 minutes for assessing knowledge. The researchers then started to assess care provided by mothers during their actual work at therapy session during hydrocortisone injection The researchers were available by rotation 3 days per week: Sunday in Benha University Hospital, Monday in the Specialized Pediatric Hospital and Thursday in Health Insurance Hospital. At the beginning of the first session, an orientation of the contents was listed and then explanation, demonstration and re demonstration were done. After finishing data collection the actual mothers care that given for their children

Procedure:

- **Preparation phase:**

It was concerned with designing and testing different data collection tools, in addition, the administrative arrangements to carry out the study as well as to conduct the pilot study. In the beginning, the researchers introduce themselves to the mothers. Mothers who accept to participate in the study individually interviewed by the researchers to explain the nature, purposes, and the desired outcomes of the study and an oral consent was obtained from these mothers.

Implementing phase:

Researchers asked each mother while being interviewing to feel free and not restricted or embarrassed when answering any questions, research investigators started to collect socio demographic data, followed by asking mothers about their knowledge about congenital adrenal hyperplasia (CAH) which took about 15-20 minutes. Researchers started first to collect data from mothers of children, first collect socio demographic data of mothers which took about 10 minutes then research investigators asked them about congenital adrenal hyperplasia (CAH). The researchers clarified and answered any related questions. Then, each mother was observed during their practice using mothers` observational check list by the same researcher. The time needed for each observation for each mother was 20-25 minutes.

Evaluation Phase:

The effect of the educational program on the study subjects was carried out through comparing the pre and post assessment score of mothers` knowledge, practice and quality of life of children.

Administrative design

An official permission for data collection was obtained from the hospitals' managers through submission of official letters issued from the dean of Benha faculty of nursing. The title, objectives, and outcomes of the study were illustrated as well as the main data items to be covered, and the study was carried out after gaining the necessary permission. The study was carried out during the period from beginning of January 2018 to the end of December 20 18.

Statistical design

The collected data revised, organized, tabulated and analyzed by using SPSS (Statistical Package for the social Science Software) statistical package version 20 on IBM compatible computer. Numerical data (Quantitative data) was presented in tables by using Mean, Standard deviation (X ± SD) and analyzed by applying t-test for normally distributed variables, while qualitative data were expressed as frequency and percentage and chi-square was used. Additionally, other statistical tests such as Independent t test was used as a parametric test of significance for comparison between two samples means. Pearson correlation (r) was used to measure the correlation between quantitative variables.

P-value at .05 was used to determine significance regarding:

- P-value > .05 to be statistically insignificant.
- P-value ≤ 05 to be statistically significant.
- P-value ≤ 001 to be high statistically significant.

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3. RESULTS

Table (1) reveals the characteristics of the studied mothers. The mean age of mothers was 31.96±4.99, more than one third of them (38.0%) were between 30-<35 years, less than half of them (45.0%) were having secondary school education and more than half of them (53.0%) were living in rural residence.

Table (2) shows the characteristics of the studied children. The mean age of the children was 6.72±3.49 years, slightly more than half of them (51.0%) were males, almost less than half (40.0%) were in primary school and more than half of them (53.0%) were ranked as the first child in the family.

Table (3) reveals that the family and illness history of the studied children with congenital adrenal hyperplasia. Less than two third of the children (60.0%) having consanguinity relation between the father and the mother, almost three quarters of them (70.0%) reported that no positive family history of congenital adrenal hyperplasia and more than half of them (54.0%) take the injection of hydrocortisone two times/ month.

Table (4) shows that, there was a highly statistically significant difference in knowledge score of the studied mothers` regarding to congenital adrenal hyperplasia at post-program as compared to pre-program (P=<0.001).

Table (5): shows that there was a highly statistically significant difference in mothers` reported practice regarding to care of congenital adrenal hyperplasia at post of educational implementation as compared to pre of program implementation (P=<0.001).

Table (6) shows that there was a highly statistically significant difference observed between the studied mothers according to the quality of life domains at pre/ post of program implementation.

Table (7) shows that there was a highly statistically significant difference observed between the studied mothers` total knowledge, quality of life and mothers` reported practice regarding to congenital adrenal hyperplasia at pre and post of educational implementation P= (<0.001).

Table (8): Revealed that, there were positive correlation between personal characteristics of studied mothers` and total knowledge, quality of life and practice (p value < .001) after educational program implementation

Figure (1) shows that there was a highly statistically significant difference observed between the studied mother's total knowledge score regarding to congenital adrenal hyperplasia at pre and post of educational intervention implementation P= (<0.001). This meaning that, one third of studied mother`s (33.0%) had average level of knowledge score regarding to congenital adrenal hyperplasia and less than one third of them (31.0%) had poor level of knowledge score at pre of educational intervention. While more than two third of them (69.0%) reported good level of knowledge score at post of educational intervention implementation.

Figure (2) shows that there was a highly statistically significant difference observed between the studied mother's total report practice score regarding to congenital adrenal hyperplasia at pre and post of educational intervention implementation P= (<0.001). This meaning that, more than half of studied mother`s (55.0%) had poor level of report practice score regarding to congenital adrenal hyperplasia at pre. While more than three quarter of them (74.0%) had good level of report practice score at post of educational intervention implementation

Figure (3) shows that there was a highly statistically significant difference observed between the studied mother's total quality of life score regarding to congenital adrenal hyperplasia at pre and post of educational intervention implementation P= (<0.001). This meaning that, had poor total score quality of life at pre of educational intervention, less than two thirds of them (63.0%) had good total score quality of life at post of educational intervention.

Table (1): Number and percentage distribution of the studied mothers` according to their personal characteristics (n=55).

Items	No	%
Age in years		
▪ 25 - <30	19	35.0
▪ 30 - <35	21	38.0
▪ 35 - <40	11	20.0

Novelty Journals

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▪ 40- ≤45	4	7.0
± SD	31.96±4.99	
Educational Level		
▪ Read and Write	2	4.0
▪ Primary	5	9.0
▪ Preparatory	15	28.0
▪ Secondary	25	45.0
▪ University	8	14.0
Residence		
▪ Urban	26	47.0
▪ Rural	29	53.0

Table (2): Number and percentage distribution of the studied children according to their personal characteristics (n=55).

Items	No	%
Age in years		
▪ <1	5	10.0
▪ 1- <6	17	31.0
▪ 6- <10	25	45.0
▪ 10- ≤16	8	14.0
± SD	6.72± 3.49	
Gender		
▪ Males	28	51.0
▪ Females	27	49.0
Educational Level		
▪ Primary	22	40.0
▪ Preparatory	25	46.0
▪ Secondary	8	14.0
Child's rank		
▪ First	29	53.0
▪ Second	16	29.0
▪ Third	5	9.0
▪ Fourth	5	9.0

Table (3): Number and percentage distribution of the studied children according to their family and illness history (n=55).

Items	No	%
Consanguinity relation between the father and the mother		
▪ Yes	33	60.0
▪ No	22	40.0
Presence of family members relatives suffering form congenital adrenal hyperplasia		
▪ No	38	70.0
▪ Yes	17	30.0
The relation in case of yes (n=39)		
▪ Sibling	8	20.5
▪ Uncles / uncles	19	48.7
▪ Others	12	30.8
Number of injectable hydrocortisone / months		
Once / m	15	28.0
Two / m	30	54.0
Three/ m	8	14.0
fours / m	2	4.0

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Table (4): Mean and standard deviation of the studied mothers' knowledge score regarding to congenital adrenal hyperplasia at pre/ post of program implementation (n=55).

Items	Pre	post	Paired t test	P
	$\overline{X}$ ±SD	$\overline{X}$ ±SD		
Definition of congenital adrenal hyperplasia	1.17±0.34	2.01±0.12	23.79	<0.001
Definition of adrenal Crisis	1.08±0.28	1.91±0.25	47.98	<0.001
Causes of CAH	1.31±0.45	2.00±0.11	15.16	<0.001
Signs and symptoms of CAH	1.03±0.17	2.00±0.00	56.57	<0.001
Treatment of congenital adrenal Hyperplasia	1.12±0.00	2.96±0.27	82.47	<0.001
Rout of administration hydrocortisone	1.95±0.21	3.00±0.00	47.93	<0.001
Sid effect of over dose hydrocortisone	1.00±0.00	2.94±0.23	81.27	<0.001
Sid effect of dose hydrocortisone is not sufficient	1.00±0.00	2.94±0.23	81.27	<0.001

Table (5): Mean and standard deviation of the studied mothers reported practice regarding to congenital adrenal hyperplasia at pre/ post of program implementation (n=55).

Items	Pre	post	Paired t test	P
	$\overline{X}$ ±SD	$\overline{X}$ ±SD		
Administer oral steroids	3.91±1.21	6.21±1.21	11,10	<0.001
Measuring fever	6.80±1.04	10..25±1.14	14.33	<0.001
Measuring pulse rate	2.61±0.29	4..25±0.84	17.12	<0.001
Measuring height	3.32±1.28	8.22±1.28	12.47	<0.001
Measuring, weight	2.22±1.28	5.22±1.28	18.93	<0.001

Table (6)

Items	Study group(No.55)				Paired(t) test	P value
	Pre-program		Post- program			
	No.	%	No.	%		
Total knowledge score						
Good	6	11.0	38	69.0	117.31	<0.001
Average	18	33.0	10	19.0		
Poor	31	56.0	7	12.0		
Total practice score						
Good	8	14.0	41	74.0	118.22	<0.001
Average	18	33.0	9	16.0		
Poor	29	53.0	5	10.0		
Total quality score						
Good	7	13.0	35	63.0	116.31	<0.001
Average	11	20.0	15	27.0		
Poor	37	67.0	5	10.0		

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Table (7) Total knowledge, quality of life and reported practice scores of studied mothers regarding congenital adrenal hyperplesis through the implementation of educational program (n=55)

Quality of life domains	Pre -Program Mean ± SD	Post -Program Mean ± SD	Paired t. test	P value
Physical Wellbeing	9.21 ± 2.39	15.75 ± 2.22	12.53	<0.001
psychological & mental wellbeing domains	12.81 ± 3.13	23.42 ± 1.52	15.60	<0.001
Social wellbeing domains	13.78 ± 3.68	22.72 ± 3.32	10.42	<0.001
Medication effect domains	7.15 ± 2.09	10.27 ± 1.008	7.40	<0.001
Total	42.95±11.29	72. 16 ± 8.068	20.31	<0.001

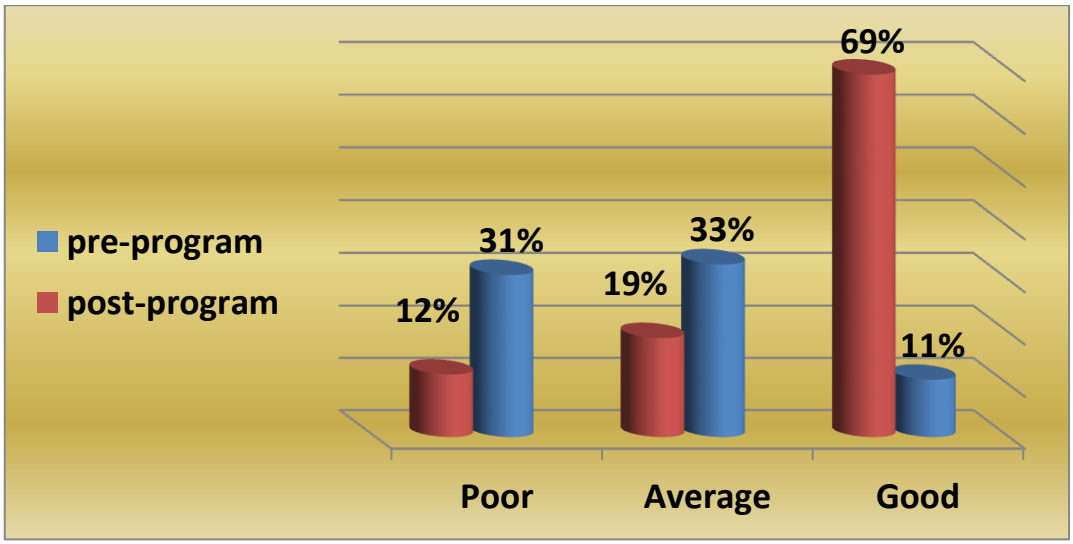

Fig. (1): Distribution of the studied mothers` regarding their total knowledge scores regarding congenital adrenal hyperplasia for children pre/post- program implementation (n=55)

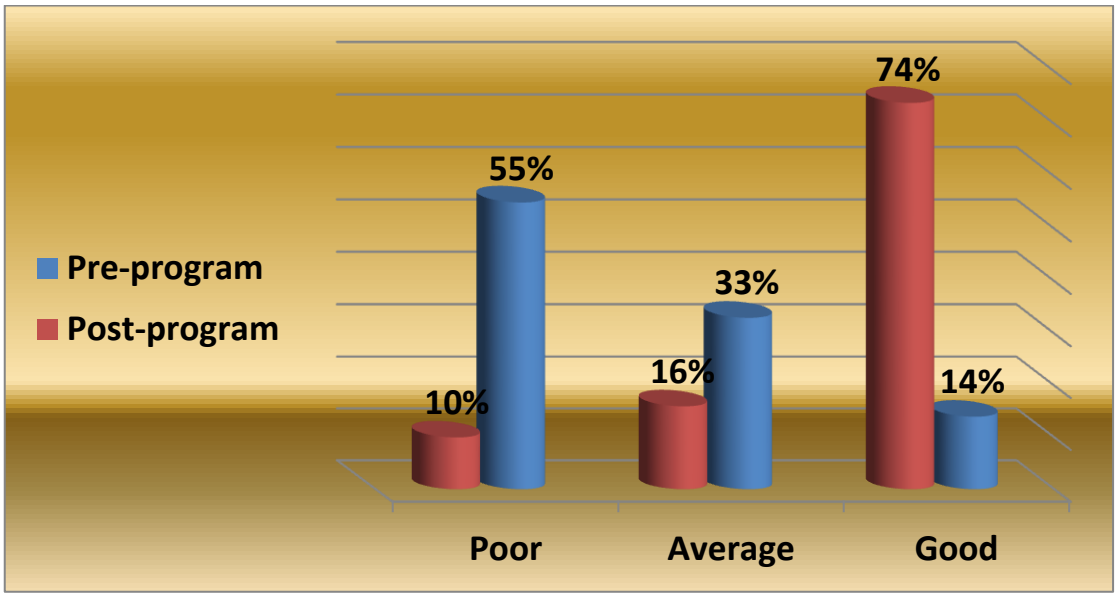

Fig. (2): Distribution of the studied mothers` regarding their total report practices scores regarding congenital adrenal hyperplasia for children pre/post- program implementation (n=55)

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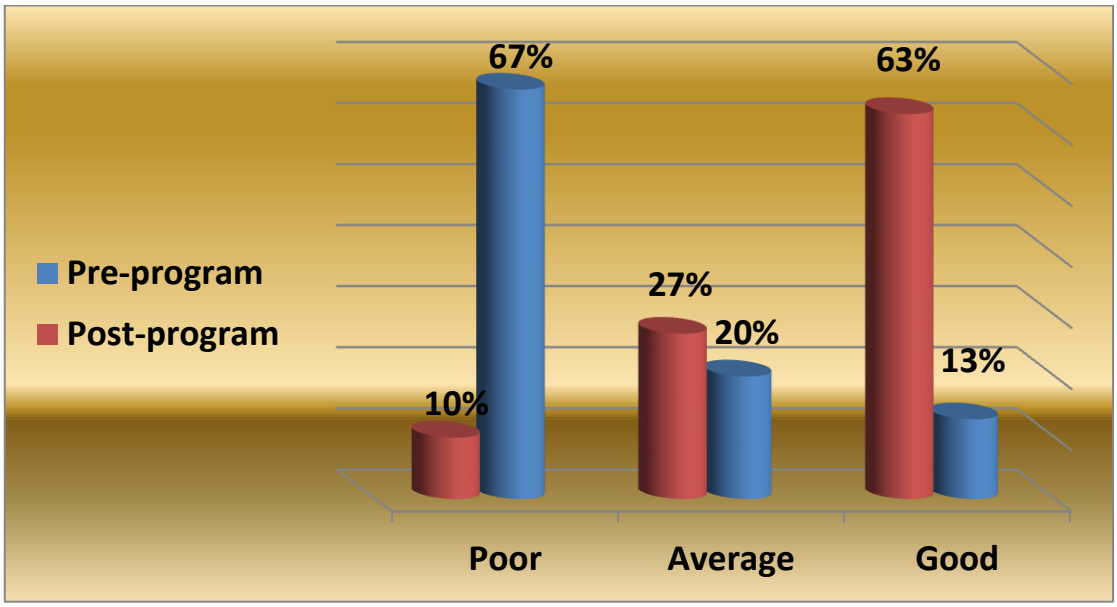

Figure (3): Distribution of the studied mothers `regarding their total quality of life scores towards children suffering from congenital adrenal hyperplasia pre/post- program implementation (n=55)

Table (8) Correlation between personal characteristics of studied mother`s and total knowledge, quality of life and practice after educational program implementation

Items	Pearson correlation coefficient					
	Knowledge score		Quality of life score		Reported practice score	
	Pearson	Sig	Pearson	Sig	Pearson	Sig
Age	.415	<.001	.387	<.001	.440	<.001
Level of educational	.529	<.001	.517	<.001	.573	<.001

4. DISCUSSION

Congenital adrenal hyperplasia (CAH) is a group of rare inherited autosomal recessive disorders characterized by a deficiency of one of the enzymes needed to make specific hormones. CAH effects the adrenal glands located at the top of each kidney. (**Speiser et al 2010**). Children with CAH are steroid dependent for life, and the goal of daily maintenance treatment is to replace deficient levels of cortisol and/or aldosterone while minimizing androgen excess, preventing virilization, optimizing growth, and protecting fertility. (**Moreira et al 2013).** This study aimed to evaluate the effect of the educational program on quality of life for mothers of children suffering from congenital adrenal hyperplasia.

Regarding to the personal characteristics of the studied mother`s, the results of the current study showed that, more than one third of them were 30-35 years with mean age 31.96±4.99 years. This result was similar to the result of a study by **Boyse et al., (2014**), study that entitled (Parents' needs after infant diagnosis with congenital adrenal hyperplasia), who found that, the age of the mothers was 30-35 years.

According to educational level of the studied mothers, the finding of the current study revealed that less than half (45.0%) were secondary school, This result is contrast with, **Abdel-Salam and Mahmoud.,** ***(2018),*** which study entitled (Effect of Educational Program on The self-Efficacy and Quality of Life for Mothers Caring children With Congenital Heart Disease) who found that, less than three quarters of the mothers' are intermediate education.

Regarding to the personal characteristics of the studied children, the results of the current study showed that, less than half of them their age ranged 6-11 years and the mean age was 6.72±3.49 years. This result was similar to the result of a study by **Pasterski et al., (2015),** entitled (Increased cross-gender identification independent of gender role behavior in girls with congenital adrenal hyperplasia, who found that age from 4- to 11-year-old children.

According to gender of the studied children, the results of the current study showed that, more than half of them were males. This result was similar to the result of study by, **de Silva et al., (2011)**, entitled (Psychological impact on parents of children with congenital adrenal hyperplasia), who found that, half of children (50.0%) were males.

According to level of education of the studied children results of the current study showed that, less than half were in primary school. This finding is similar with **Berenbaum et al., (2012)**, entitled (Early androgen effects on spatial and mechanical abilities: evidence from congenital adrenal hyperplasia) who found that less than half of children were in primary school.

Regarding to the rank of the child in the family, the findings of the current study showed that, more than half were ranked as the first child of the family. This finding is similar with **Boyse et al., (2014),** who found that, (52.3%) of the studied children was ranked as the first child of the family. This could reveal that, the mothers with a first child suffering from congenital adrenal hyperplasia may have a lack of knowledge and experiences regarding to CAH disease due to lake aware of them for this disease.

The present study has also revealed that mothers' knowledge about definition of congenital adrenal hyperplasia, definition of adrenal crisis, causes of CAH, signs and symptoms of CAH, treatment of congenital adrenal hyperplasia, rout of administration hydrocortisone, side effect of over dose hydrocortisone and side effect of dose hydrocortisone is poor in the pre-program phase, but it improved in the post-program implementation phase, and there were a highly statistical significant difference (P< 0.001) regarding total knowledge scores, this applied to all related areas of knowledge. On the same line **Kim et al., (2012),** study entitled; (Management of congenital adrenal hyperplasia in childhood. Current Opinion in Endocrinology, Diabetes, and Obesity),who found that most of the parents of children with CAH were having a lack of knowledge in relation to symptoms and treatment of congenital adrenal hyperplasia and a lack of medical home and decision support

The current study assessed mother`s reported practice toward caring of their children suffering CAH, the study findings demonstrated poor levels of practice among them before implementation of the program. This was particularly evident in crucial tasks such as administer oral steroids, measuring pulse rate, measuring weight, measuring height and measuring fever . On the same line **Mendes-Dos-Santos(2011)**, in study entitled; (Normalization of height and excess body fat in children with salt-wasting 21-hydroxylase deficiency), who found that most of the parents of children with CAH have very low levels of practice and a lack of medical home and decision support. This may be due to lack of training and information of mothers about the impact of the disease and treatment on their children, in response to this problem,

On the other hand as mentioned by **(Louise et al 2011),** 21- hydroxylase enzyme deficiency is the most common cause of CAH which is characterized by cortisol deficiency, with or without aldosterone deficiency, and androgen excess.

Concerning the reported practice of the mothers about congenital adrenal hyperplasia, the current study revealed that, improvement in their reported practice regarding to administer oral steroids, measuring pulse rate, measuring weight, measuring height and measuring fever after implement of the educational intervention and there were a highly statistical significant difference (P< 0.001) regarding total reported practice scores, .This result can be explained in the light of **Sanches et al (2012)**, entitled; (Physical, social and societal functioning of children with congenital adrenal hyperplasia (CAH) and their parents, in a (dutch population),who found that mother`s guideline and a demonstration on how to give the hydrocortisone injection and oral medication scored higher.

Regarding to the domain quality of life for children with congenital adrenal hyperplasia (CAH), the current study found that, more than two third of the studied children had poor quality of life level regarding to physical status and total psychological & mental wellbeing domains, total Social wellbeing domains and total medication effect domains at pre of educational program. These findings agree with **Gilban et al (2014),** entitled; Health related quality of life of children and adolescents with congenital adrenal hyperplasia in brazil. Health and Quality of Life Outcomes), who found a loss of health related quality of life in children and adolescents with classical CAH.

Regarding to all domains of quality of life for those children, the current study showed that, improving the quality of life for children in all domains after implementing the educational intervention for them as compared to pre educational intervention .These findings is in an agreement with, **Borhani et al., (2011), entitled** (The effect of Family-Centered Empowerment Model on Quality of Life of School–Aged Children with Thalassemia Major), who found that, increasing

quality of life of children in all domains after implementing the family-centered empowerment model. Also **Gilban et al (2014)** , found that health related quality of life of children and adolescents with CAH can improve can the concerning measures of parents and children to prevent an adrenal crisis.

Regarding to the correlation between total knowledge , report practices of the mothers and their total quality of life, the current study revealed a positive correlation between mother`s knowledge, report practices and their total quality of life . This result is a supported with **AL-Mosooi, (2011)**, in a study entitled (Assessment of Health Quality of Life upon School Age Children (6-12) with Thalassemia), who found that, there was a statistical significant association between knowledge of the school age children with thalassemia and their all quality of life domains.

The educational program for mother`s with congenital adrenal hyperplasia (CAH) can be achieve a higher knowledge and report practices and would improve the ability of the mother`s in health decision-making and consequently would cause changes in their behavior,

5. CONCLUSION

The educational program was effective in improving the mothers' knowledge and reported practice regarding congenital adrenal hyperplasia in all dimensions of the quality of life for the children with congenital adrenal hyperplasia: Physical, social, psychological & mental wellbeing domains, and total medication effect domains

6. RECOMMENDATIONS

Based on results of the present study, it can be recommended that;

- The developed program should be applied and repeated again every 6 months in the same study settings and adopted in other similar settings with required modifications.

- provision of continuing education programs on regular basis is suggested in order to refresh and update mother`s knowledge, as well as reinforce proper practices related to congenital adrenal hyperplasia in pediatric units

- Understanding and identifying gaps in parent education by healthcare professionals, especially concerning times of adrenal crisis.

- Mothers' wrong practice about their children with congenital adrenal hyperplasia compliance related to giving preventive measure and the regularity of follow-up for the child's condition should be corrected through health teaching.

Future studies;

- An emphasis on family experience and management would enhance the current state of the science and provide a much-needed window into interventions aimed at improving the lifes of families and children with CAH.

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