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Level of Nurses' Performance Regarding Care of Infants with Protein C Deficiency: Intervention Study

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Abstract: **Protein C deficiency is one of the major inhibitors of the coagulation system and associated with an increased risk of venous thrombosis. The aim of this study was to evaluate the effect of the intervention study for nurse's performance regarding care of infants with protein C deficiency. A quasi-experimental design was used. This study was carried out at Benha specialized hospital of for Children affiliated to Ministry of Health. A convenient sample included all nurses (50), who are working at Benha Specialized Hospital the Children. Tools of data collection: Two tools were used; Tool I: A structured interviewing questionnaire, which consists of four parts: Part (1) Nurses' characteristics, Part (2): Nurses' knowledge about Protein C deficiency (PCD),Part (3): Nurses` knowledge regarding Protein C and its function, Part (4): Nurses` knowledge regarding management, Tool II: Nurses' practice sheet. Results indicated a highly statistically significant difference observed between the studied nurses total knowledge and practice regarding Protein C deficiency at pre and post of the intervention implementation P= (<0.001)..This study concluded that: after implementation intervention study of the nurses had satisfactory knowledge and competent practice, than before implementation regarding protein C deficiency for children. The study recommended that: future studies should emphasis on nurses' experience and management would enhance the current state of the science and provide a much-needed window into interventions aimed to improving the medical outcomes for children with protein C deficiency.**

Keywords: **Protein C deficiency, Infant's, Level of Performance & Intervention Study.**

1. INTRODUCTION

Protein C is an anticoagulant factor that is normally present in blood. It has anti-inflammatory effects on endothelial cells and leukocytes .APC affects endothelial cells by inhibiting inflammatory mediator release and down-regulating vascular adhesion molecules. This reduces leukocyte adhesion and infiltration into tissues, while, it also limiting damage to underlying tissue **(Zia et al.,2015)**.

Protein C deficiency is an inherited disease results in lower than normal level of protein C in the blood. It is rare, occurring in about 1 in 500 people in the United States. Protein C deficiency is associated with an increased incidence of venous thromboembolism, whereas no association with arterial thrombotic disease has been found. It is rare but is a life-threatening disorder that can optimize outcomes for these unique infants of coagulation. Some complications may be inevitable, but prompt recognition and treatment of the underlying disease (**Nicolaes etal., 2014)**.

Clinical manifestations appear in the form of severe genetic protein C deficiency which usually presents within hours of birth with rapidly progressive purpura fulminant (PF) and disseminated intravascular coagulation (DIC. PF originates with red or purpuric lesions at pressure points, such as the back of the head and buttocks.The lesions rapidly progress to form palpable black and children with severe protein C deficiency experience recurrent episodes of PF triggered by infection,

trauma, were as most common symptom is deep vein thrombosis. This is a clot that forms in the deep veins of the legs. This can be painful and can cause the leg to swell, but the clots can form without pain or swelling too (**Rao,.2009).**

Protein C deficiency is divided into Type I or Type II deficiency. Type 1: caused by low levels of normal protein C and, Type 2: caused by an abnormally functioning protein **(Taiaku et al 2015).** Most infants who have have no symptoms or simply have an increased risk for deep vein thrombosis during recovery from surgery or periods of immobility.Children with the milder form of protein C deficiency may not show any symptoms (asymptomatic) until they reach adulthood. Others may remain asymptomatic(**Manco-Johnson et al., 2016**).

Complications of this disease include ophthalmic problems, central nervous system (CNS) and pulmonary embolism. Blood clots can also form in the blood vessels that drain blood from the large and small intestines (mesenteric veins). Less often, blood clots may form in the cerebral veins, the main vein of the liver (portal vein), and other areas**(Bauer 2016).**

Management consists of correcting the coagulopathy, intensive wound care including negative-pressure dressings and skin grafting and supportive care for the ophthalmic and CNS issues. A most important element of wound healing is providing excellent nutrition. Poor nutrition in children with significant wounds can extend the healing process by "prolonging the inflammatory phase, decreasing fibroblast proliferation, and altering collagen synthesis," as well as by increasing chances of infection. Multivitamins are recommended, with extra amounts of vitamins A and C because these enhance collagen deposition and fibroblast growth **(Chalmers et al 2011**)

Significance of study:

Protein C (PC) deficiency is a rare but life-threatening bleeding disorder that can present in infants and children. Whereas, with protein C deficiency have inherited the abnormal protein C gene from one parent. There is a very rare chance (less than one in a million) of a child being born with two copies of an abnormal protein C gene, one from each parent. This condition causes severe problems with blood clotting leading to skin ulcers, blindness and organ failure shortly after birth. **(Gavva et al., 2017**)The incidence of protein C deficiency in children who present with clinical symptoms has been reported to be estimated at 1 in 20,000. (**Chalmers et al 2011)**.The researchers observed that infants with protein C deficiency were at high risk for many complications related to the lack of knowledge and incompetent practice given to those infants. Hence, the present study is under taken to evaluate the effect of the intervention study for nurse's performance regarding care of infants with protein C deficiency.

Aim of the study:

The aim of this study was to evaluate the effect of the intervention study for nurse's performance regarding care of infants with protein C deficiency.

Research Hypotheses:

Nurses` performance regarding care of infants with protein C deficiency significantly will be improved after intervention study

2. SUBJECTS AND METHOD

Research Design:

A quasi- experimental research design was utilized.

Settings:

The study was carried out at Benha Specialized Hospital for Children affiliated to Ministry of Health in the neonatal intensive care unit and neonatal surgical unit

Subjects:

A convenience sample included all the available nurses (50), who were working at the previously mentioned setting

Tools of Data Collection:

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

Tool I: A structured interviewing schedule: It was developed by the researchers after reviewing the related literatures (**Nicolaes, et al., 2010, Bauer2016, and Tcheng,et al.,2008)** and it was written in Arabic language to suit the study sample. It composed of four parts as following:

Part (1): Nurses' characteristics, such as; age, gender, Level of education, years of experience.

Part (2): Nurses' knowledge about Protein C deficiency (PCD) which includes; definition, the presenting features of PC, diagnosis, predisposing factors, degrees, common kinds of symptoms, subtypes of PC deficiency, clinical symptoms, treatment, complications, laboratory investigations and admission lab tests, long-term outcomes, Lifelong therapy and prognosis.

The total questions were (13 questions) and in a form of multiple choice questions.

Part (3): Nurses` knowledge regarding Protein C and its function such as; definition of PC, normal ranges of PC, functions of PC, importance of activated PC and side effects accompanied with Activated PC.

The total questions were 5 questions) and in a form of multiple choice questions.

Part (4): Nurses` knowledge regarding about management for children with PC deficiency, such as; correcting the coagulopathy with PC replacement, intensive wound care, parenteral nutrition (PN) with protein and calories as tolerated , supportive care for the ophthalmic functions and follow up, prevention of infection and follow up and parental education.

The total questions were (6 questions) and in a form of multiple choice questions.

Scoring system for nurses` knowledge:

For each knowledge items, The scoring system consisted of giving score (2) for the complete correct response, while the correct incomplete response was scored (1); and don't know or incorrect response was scored (0).Total Scoring was classified as less than (75%) considered unsatisfactory, while more than(75%) considered satisfactory

Tool II: Observational checklist for nurses' practices

It was adopted from **Chalmers et al.,(2011)** to assess nurses' practice regarding care of children with PC deficiency. It included; intensive wound care, correcting the coagulopathy, pain management and supportive care for the ophthalmic issue.

The total practices were 10 practices which included (28 items). It included the following items:

1- -Intensive wound care (9 items)

2- Correcting the coagulopathy (7items)

3- -Pain management. (8 items)

4- Supportive care for the ophthalmic issue. (4 items)

-.**Scoring system for nurses` practices was as follows :**

Done correctly was scored (2)

Done incorrectly was scored (1)

Not done was scored (zero)

The total score of practice' steps responses was 100%, accordingly more than 80%was considered competent, and less than 80% was considered incompetent.

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 blood diseases department 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 clearer and more precise. The suggested changes were made. Internal consistency reliability of all items of the tools was assessed using Chronbach's Alpha test. It was 0.83for the structured interviewing schedule, and 0.86 for nurses' observation checklist.

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 nurse was informed about the purpose, procedure, benefits, and nature of the study and each nurse informed that she 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 nurses) 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 25 minutes.

Field of Work:

Data were collected from the beginning of September 2018 to the end of February 2019. Immediately after the ethical approval was obtained; the researchers obtained oral consents from nurses 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 nurse individually and this took about 20-25 minutes for assessing knowledge. The researchers then started to assess care provided by nurses during their care for each infant 3days / week with follow up for infants` progress condition before and after care. The researchers were available by 3 days per week: Sunday, Monday and Tuesday in the Specialized Pediatric 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 nursing care was assessed and practices applied to nurses about care for infants.

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 introduced themselves to the nurses. Nurses 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 nurses.

I. Assessment phase:

It was carried out using tool one & two to collect baseline data and to detect nurses' knowledge/practice needs.

II. Planning phase:

The intervention study was planned based on assessment phase and recent review of literature. It included goals & contents.

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Each session was started by a summary about what has been discussed in the previous one and presenting the objectives of the new session, using simple Arabic language, also the session ended by a summary of its content and feedback from the nurses was obtained to ensure that they got the maximum benefit.

Implementation Phase:

The intervention study was conducted through six sessions; theoretical part was performed through lectures and group discussions which were done in 2 sessions for theoretical, (each session lasted for 20-25 minutes) according to the nurses' needs and condition in groups (place and time). At the beginning of the first session, the intervention content and its aim were explained to the nurses. Then conduction of practical part began at the previously mentioned settings. It was taken 4sessions (each session for one hour) and covers the intervention items.

-The data were collected during a period of 6 months from the beginning of September 2018 to the end of February 2019.

- Each nurse was interviewed individually by the researcher. The mean time needed for each sheet was about 20-25 minutes to complete a questionnaire.

This study was carried out in five separate steps: developing an interview questionnaire sheet, pilot study, assessment of baseline nurses' knowledge and practical (pre-test), implementation of intervention study and evaluation of nurses' knowledge and practice after intervention study.

Each session was started by a summary about what has been conversed in the previous one and presenting the objectives of the new session using simple Arabic language, also the session was finished by a summary of its content and feedback was gained to ensure that they got the maximal benefit.

The total number of the subjects was 50 nurses; they were divided into 10 groups, 5 nurses in each group. The intervention was presented to each group separately.

Different teaching strategies were used for implementation of the intervention study such as lectures, small group discussion, brain storming, role play, demonstration and re-demonstration using real objects. Suitable teaching aids as booklet, colored posters, doll and real objects were prepared especially for practice. Nurses were motivated to cooperate and participate actively in different stages of the study.

The intervention study was carried out in the following six sessions

First session contents were (the aim of the intervention, importance, Their Knowledge about Protein C Deficiency (PCD), its function (definition, the presenting features of PC, diagnosis, Predisposing factors, Degrees, common kinds of symptoms, Subtypes of PC deficiency, Clinical symptoms, Treatment, Complications, Laboratory investigations and admission lab tests, long-term outcomes, Lifelong therapy and Prognosis).

Second session about management of PCD contents were (correcting the coagulopathy with PC replacement, intensive wound care, Parenteral nutrition (PN) with protein and calories as tolerated , supportive care for the ophthalmic functions and follow up, prevention of infection and follow up and parental education) .

Third session contents were practice in relation to(intensive wound care

Wound assessment, wound cleansing, wound dressing and follow dressing grid)

Fourth session contents were correcting the coagulopathy (administration of fresh frozen plasma (FFP) and Protein C replacement).

Five session contents were Pain management

Six session contents were supportive care for the ophthalmic issue (Daily eye care, regular retinal observation for bleeding and Long-term follow-up education for lifelong anticoagulant therapy)

VI. Intervention study evaluation:

The effect of the program on the study subjects was carried out through comparing the pre and posttest immediately assessment score of nurses knowledge& practice

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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 September 2018 to the beginning of February 2019

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 b

3. RESULTS

Table (1) revealed the personal characteristics of the studied nurses whereas, less than one thirds (32.0%) of the them were in the age category 30<35years with mean age (31.7±7.5). While, the majority of them (84%) were females. Regarding their level of education, it is obvious that, nearly two thirds (62.0%) of them had technical nursing institute was the highest proportion. More than two fifth of them(42.0%) had years of experience 5>10year years with mean years of experience (7.6±5.2).

Table(2),demonstrated that the distribution of the studied nurses according to their knowledge about PCD .Whereas, nearly three quarters (70%, %70%,%74) respectively had incorrect answers regarding clinical symptoms, prognosis and treatment on pre- intervention phase compared with post - intervention ,with statistical significant differences before and after intervention

Table(3) illustrated that the distribution of the studied nurses according to their knowledge about Protein C .Whereas, nearly two thirds (66%,,66%,68%) respectively had incorrect answers regarding definition of PC, side effects accompanied with activated PC and importance of activated PC on pre- **intervention** phase compared with post-intervention ,with statistical significant differences before and after intervention

Table (4) demonstrated that the distribution of the studied nurses according to their knowledge about management of Protein C deficiency. Whereas, more than two thirds (66%,68%,68%) respectively had incorrect answers regarding Correcting the coagulopathy with PC replacement, Supportive care for the ophthalmic functions and follow up and follow up and parental education on pre- intervention phase compared with post- intervention i ,with statistical significant differences before and after intervention .

Table (5) revealed that the distribution of the studied nurses according to their practice in relation to caring for children with PCD. Whereas, there was a progressive improvement in their practice on pre- intervention phase compared with post intervention, with statistical significant differences before and after intervention

Table (6), revealed that, there were a statistically significant relation between personal characteristics of the studied nurses and their total knowledge scores pre intervention. While, there was no statistically significant correlation post intervention.

Table (7) revealed that there was a statistically significant relation between personal characteristics of the studied nurses and their total practices scores post intervention study compared with pre intervention.

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Table (8) indicated that a statistically significant positive correlation between nurses` total knowledge scores before intervention and their practice after intervention

(Table (1): Number and percentage distribution of the studied nurses according to their personal data (No= 50)

Items	No	%
Nurses' age		
20<25	15	30.0
25<30	10	20.0
30<35	16	32.0
≥35	9	18
Mean ±SD	**7.5±31.7**	
Gender		
Male	8	16.0
Female	42	84.0
Level of education		
Diploma (secondary school)	9	18.0
Technical nursing institute	31	62.0
Bachelor degree	10	20.0.
Years of experience		
1<5year	17	34.0
5>10year	21	42.0
≥10year	12	24.0
Mean ±SD	**5.2±7.6**	

Table (2): Number and Percentage distribution of the studied Nurses regarding their knowledge about Protein C deficiency (PCD) pre and post intervention. (No=50)

Items	Pre intervention						Post intervention						^2X	p-value
	Complete Correct answer		incomplete Correct answer		Incorrect or answer Don't know		Complete Correct answer		incomplete Correct answer		Incorrect or answer Don't know			
	no	%	no	%	no	%	no	%	no	%	no	%		
-Definition of protein C deficiency	4	8.0	13	26.0	33	66.0	41	82.0	8	16.0	1	2.0	53.8	0.000
- Predisposing factors	2	4.0	18	36.0	30	60.0	35	70.0	15	30.0	0	0.0	59.7	0.000
-The presenting features of PC deficiency	5	10.0	11	22.0	34	68.0	33	66.0	12	24.0	5	10.0	42.2	0.000
-Degrees	4	8.0	13	26.0	33	66.0	37	74.0	9	18.0	4	8.0	50.0	0.000
-Common kinds of symptoms	8	16.0	13	26.0	29	58.0	35	70.0	11	22.0	4	8.0	36.0	0.000
-Subtypes of PC deficiency	4	8.0	14	28.0	32	64.0	39	78.0	9	18.0	2	4.0	56.0	0.000

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Table (3): Continued, Number and Percentage distribution of the studied Nurses regarding their knowledge about Protein C deficiency (PCD) pre and post intervention. (No=50)

Items	Pre intervention						Post intervention						^2X	p-value
	Complete correct answer		incomplete correct answer		Incorrect or answer Don't know		Complete correct answer		incomplete correct answer		Incorrect or answer Don't know			
	no	%	no	%	no	%	no	%	no	%	no	%		
-Clinical symptoms	5	10.0	10	20.0	35	70.0	48	96.0	1	2.0	1	2.0	74.3	0.000
Treatment	0	0.0	13	26.0	37	74.0	38	76.0	8	16.0	4	8.0	65.7	0.000
-Complications	8	16.0	15	30.0	27	54.0	36	72.0	9	18.0	5	10.0	34.4	0.000
-Laboratory investigations and admission lab tests	8	16.0	13	26.0	29	58.0	35	70.0	11	22.0	4	8.0	36.0	0.000
-Long-term outcomes	3	6.0	19	38.0	28	56.0	41	82.0	6	12.0	3	6.0	59.7	0.000
-Lifelong therapy	8	16.0	15	30.0	27	54.0	36	72.0	9	18.0	5	10.0	34.4	0.000
-Prognosis	5	10.0	10	20.0	35	70.0	48	96.0	1	2.0	1	2.0	74.3	0.000

Table (4): Number and percentage distribution of the studied nurses regarding their knowledge regarding PC and its function (No=50)

Items	Pre intervention						Post intervention						^2X	p-value
	Complete correct answer		Incomplete correct answer		Incorrect or answer Don't know		Complete correct answer		Incomplete correct answer		Incorrect or answer Don't know			
	no	%	no	%	no	%	no	%	no	%	no	%		
Definition of PC	4	8.0	13	26.0	33	66.0	37	74.0	9	18.0	4	8.0	50.0	0.000
Normal ranges of PC	2	4.0	18	36.0	30	60.0	35	70.0	15	30.0	0	0.0	59.7	0.000
Functions of PC	6	12.0	21	42.0	23	46.0	41	82.0	9	18.0	0	0.0	53.8	0.000
Importance of Activated PC	5	10.0	11	22.0	34	68.0	33	66.0	12	24.0	5	10.0	42.2	0.000
Side effects accompanied with Activated PC	4	8.0	13	26.0	33	66.0	37	74.0	9	18.0	4	8.0	50.0	0.000

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Table (5): Number and percentage distribution of the studied nurses regarding their knowledge about management pre and post intervention (No=50)

Items	Pre intervention						Post intervention						^2X	p-value
	Complete correct answer		Incomplete correct answer		Incorrect answer or Don't know		Complete correct answer		Incomplete correct answer		Incorrect answer or Don't know			
	no	%	no	%	no	%	no	%	no	%	no	%		
Correcting the coagulopathy with PC replacement	4	8.0	13	26.0	33	66.0	37	74.0	9	18.0	4	8.0	50.0	0.000
Intensive wound care	6	12.0	21	42.0	23	46.0	41	82.0	9	18.0	0	0.0	53.8	0.000
Parenteral nutrition (PN) with protein and calories as tolerated.	2	4.0	18	36.0	30	60.0	35	70.0	15	30.0	0	0.0	59.7	0.000
Supportive care for the ophthalmic functions and follow up	5	10.0	11	22.0	34	68.0	33	66.0	12	24.0	5	10.0	42.2	0.000
Prevention of infection	6	12.0	21	42.0	23	46.0	41	82.0	9	18.0	0	0.0	53.8	0.000
follow up and parental education	5	10.0	11	22.0	34	68.0	33	66.0	12	24.0	5	10.0	42.2	0.000

Table (6): Number and percentage distribution of the studied nurses regarding their practice in relation to caring for neonates with PCD pre and post intervention (No=50)

Items	Pre intervention						Post intervention						^2X	p-value
	Done correctly		Done incorrectly		Not done		Done correctly		Done incorrectly		Not done			
	no	%	no	%	no	%	no	%	no	%	no	%		
Intensive wound care														
Wound Assessment.	24	48.0	13	26.0	13	26.0	37	74.0	6	12.0	7	14.0	7.14	0.02
Wound Cleansing	16	32.0	18	36.0	16	32.0	37	74.0	13	26.0	0	0.0	25.1	0.000
Wound dressing,	18	36.0	13	26.0	19	38.0	40	80.0	7	14.0	3	6.0	6.7	0.030
-Follow dressing grid	2	4.0	5	10.0	43	86.0	43	86.0	4	8.0	3	6.0	72.2	0.000
Correcting the coagulopathy														
administration of fresh frozen plasma (FFP)	18	36.0	13	26.0	19	38.0	40	80.0	7	14.0	3	6.0	21.7	0.000
Protein C replacement	16	32.0	18	36.0	16	32.0	37	74.0	13	26.0	0	0.0	25.1	0.000
Pain management.	12	24.0	16	32.0	22	44.0	42	84.0	7	14.0	1	2.0	3..39	0.000
supportive care for issue the ophthalmic														
Daily **eye care**.	4	8.0	4	8.0	42	84.0	41	82.0	4	8.0	5	10.0	59.5	0.000
regular retinal observation for bleeding	16	32.0	18	36.0	16	32.0	37	74.0	13	26.0	0	0.0	25.1	0.000
. Long-term follow-up education for lifelong anticoagulant therapy	18	36.0	13	26.0	19	38.0	40	80.0	7	14.0	3	6.0	21.7	0.000

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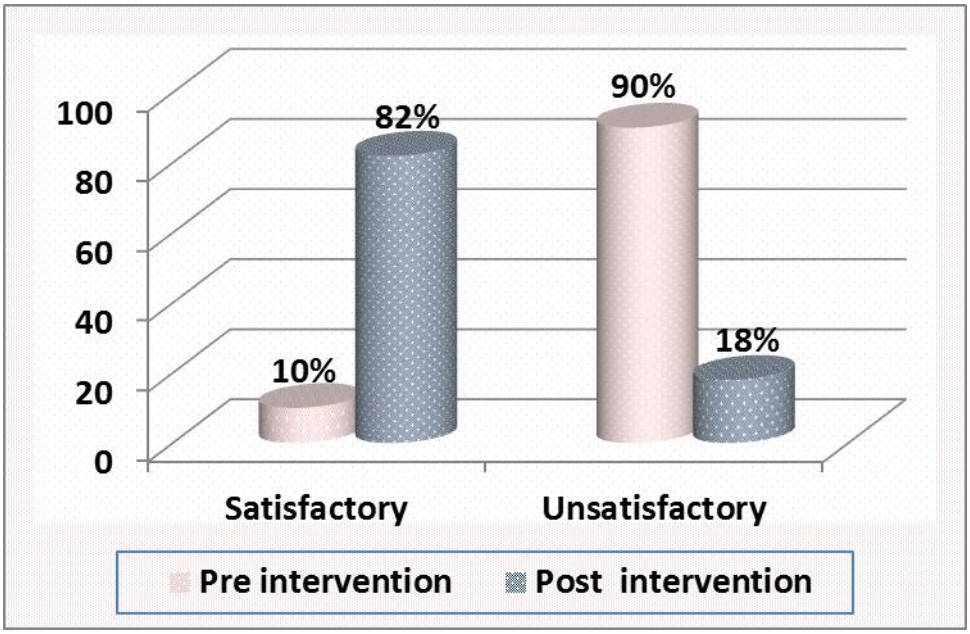
Total knowledge



Total Practice

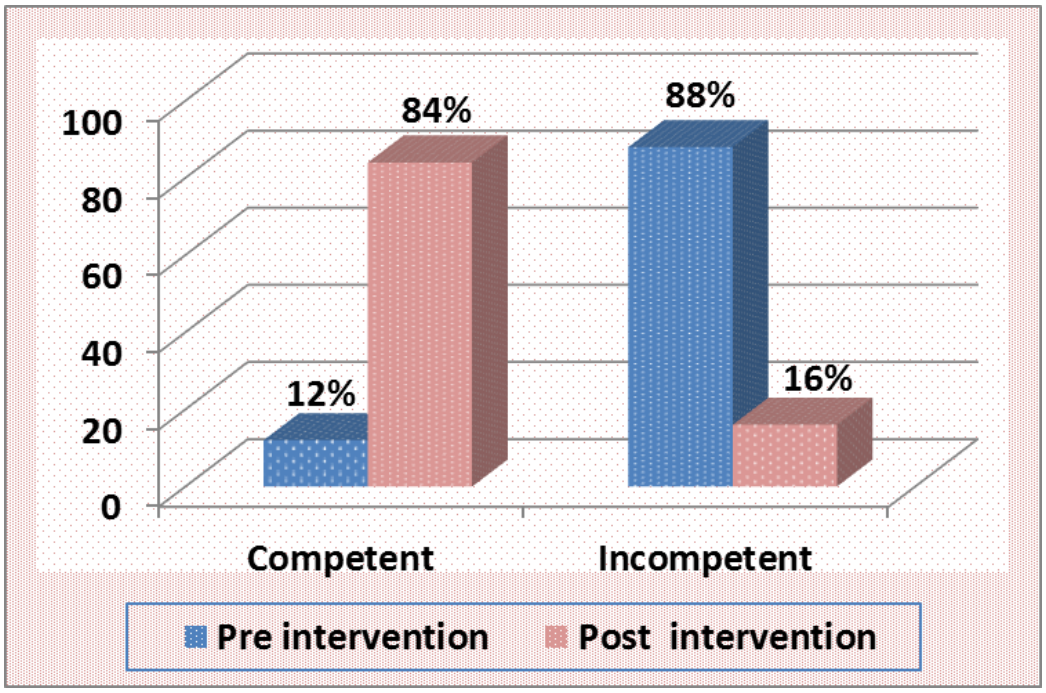


Table (7): Relation between total knowledge scores and their personal characteristics of the studied nurses pre and post intervention (No=50)

Items	Pre intervention						Post intervention					
	Complete correct		Incomplete correct		Incorrect		Complete correct		Incomplete correct		Incorrect	
	on	%	no	%	no	%	no	%	no	%	no	%
Age												
20<25	0	0.0	1	10.0	14	37.8	12	31.5	3	37.5	0	0.00

25<30	1	33.3	2	20.0	7	18.9	7	18.4	2	25.0	1	33.3
30<35	1	33.3	1	10.0	14	37.8	11	28.9	3	37.5	2	66.7
≥35	1	33.3	6	60.0	2	5.40	8	21.05	1	12.0	0	0.0
	16.52=^{2}X *p-value = 0.001						**-4.68=^{2}X p-value =0.585**					
Gender												
Male	1	33.3	1	14.20	6	15.0	5	13.50	2	20.0	1	33.3
Female	2	66.7	6	85.80	34	85.0	32	86.50	8	80.0	2	66.7
	17.30=^{2}X *p-value =0.061						**2.50=^{2}X p-value =0.123**					
Level of education												
Diploma (nursing school)	1	20.0	5	20.0	3	15.7	6	16.2	2	28.5	1	16.6
Diploma of Technical nursing institute	2	40.0	14	56.0	15	78.9	24	64.8	5	71.5	2	33.3
Bachelor degree in nursing	3	60.0	6	24.0	1	5.20	7	18.9	0	00.0	3	50.0
	***p-value =0.004- 12.6=^{2}X**						**p-value =0.106 - 9.70=^{2}X**					
Years of experience												
1<5year	1	25.0	2	15.3	14	40.0	12	31.5	3	42.8	2	50.0
5>0year	2	50.0	7	53.8	12	34.2	17	44.7	3	42.8	1	25.0
≥10year	1	25.0	4	30.7	7	20.0	9	23.6	1	14.2	1	25.0
	***p-value =0.050 -12.57=^{2}X**						**p-value =0.040 -5.59=^{2}X**					

Table (8): Relation between total practices scores and personal characteristics of the studied nurse's pre and post intervention

Items	Pre intervention				Post intervention			
	Incompetent		Competent		Incompetent		Competent	
	no	%	no	%	no	%	no	%
Age								
20<25	12	33.3	3	21.4	6	46.5	9	24.3
25<30	7	19.4	3	21.4	2	15.3	8	21.6
30<35	11	30.5	5	35.7	4	30.7	12	32.4
≥35	6	16.6	3	21.4	1	7.6	8	21.6
	0.87=^{2}X		**p-value = 0.75**		**1.37=^{2}X**		***p-value = 0.05**	
Gender								
Male	6	16.7	2	16.3	3	27.3	5	13.5
Female	30	83.3	12	85.7	10	76.7	32	85.5
	0.81=^{2}X		p-value = 0.36		0.46=^{2}X		p-value = 0.49	

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Years of experience								
1<5year	9	25.0	4	28.5	8	61.5	10	27.02
5>10year	16	44.4	5	35.7	3	23.07	17	45.9
≥10year	11	30.5	5	35.7	2	15.3	10	27.02
	10.47=^{2}X		***p-value =0.050**		**7.39=^{2}X**		***p-value =0.05**	
Level of education								
Diploma (nursing school)	9	25.0	0	0.00	2	15.3	7	18.9
Diploma of Technical nursing institute	23	63.8	4	28.5	7	53.8	24	64.8
Bachelor degree in nursing	4	11.1	10	71.5	4	30.4	6	16.2
	2.13=^{2}X		**P -value = 0. 03**		**4.12=^{2}X**		***p-value = 0.007**	

(P< 0.001) highly statistical significant differences **

Table (9): Correlation between total nurses' knowledge and practice scores pre/post intervention (No=50)

Variables		Knowledge pre	Practice pre	Knowledge post	Practice post
Knowledge pre	Pearson Correlation		.319**		*
	Sig. (2-tailed)		.004		
Practice pre	Pearson Correlation	.319**			
	Sig. (2-tailed)	.004			
Knowledge post	Pearson Correlation	.134	.024		
	Sig. (2-tailed)	.234	.835		
Practice post	Pearson Correlation	.273*	.015		
	Sig. (2-tailed)	.014	.894		

4. DISCUSSION

Protein C deficiency is a disorder that increases children`s risk to develop abnormal blood clots due to deficiency of Protein C. It is a protein in the body that prevents blood clotting. It may be inherited or acquired. Inherited deficiency of protein C can lead to familial thrombophilia (increased tendency toward thrombosis).Acquired protein C deficiency has been observed in children with acute leukemia, hepatic disease, nephritic syndrome; adrenal transplant(**Andrew & Schafer 2012)**.

The present study was focused on assessing nurses' knowledge and practice about nursing care provided to children with protein C deficiency and to design, implement and evaluate the educational program for those nurses based on the actual nurses' needs and relevant literatures. The study involved 50 nurses working in NICU at the pediatric hematology units in Benha Specialized Hospital for Children. They were mostly young age, with more than half holding Technical nursing institute.

Findings of the current study revealed that one third of the studied nurses in the pediatric units aged between 30< 35 years and their mean age of was 31.7±7.5years.This along with their low experience, lower level of nursing qualification and the new disease is expected to have an impact on their knowledge and practice which were revealed to be very low at the pre-intervention phase. In the same line with the present study finding **Tavares et al., (2015)**, who has emphasized that training program should be given in conjunction with a guided clinical practice, where nurses gain minimal knowledge regarding protein C deficiency can develop skills safely and competently. This givens support to the rationale of the present study.

In order to design intervention for improving nurses' knowledge and practice regarding nursing care provided to children with protein C deficiency, it was mandatory to assess their baseline knowledge and practice. The results of the present study revealed that nurses' knowledge before the intervention was generally unsatisfactory. This was quite clear regarding their knowledge about predisposing factors, the presenting features of PC, definition of protein C deficiency, common kinds of symptoms, clinical symptoms and treatment .The present study findings were supported by **Lai et al., (2017) and Ohga etal., (2013)**who found that the majority of nurses' knowledge was inadequate regarding definition of protein C deficiency, common kinds of symptoms, clinical symptoms and treatment .The author has emphasized that the role of the nurse is multifocal in these cases. In addition to the care giving role, the nurse must meet the need of child with protein C deficiency. She has also other roles as educator and advocate for the children, and should assess the child and or family caregivers` understanding of proposed treatment, evaluating child needs, the methods of administration of treatment and the potential risk from complication.

Moreover, **Price et al (2011)** has stressed that the oncology nurse should provide expert nursing care to maximize health, enhance quality of life relieve suffering and provided education and support to children and their family and making sure that children feels cared for and facilitate streamline transition from one treatment area to another.

The present study has also revealed that nurses' knowledge about importance of activated PC, normal ranges of PC and side effects accompanied with activated PC and were unsatisfactory in the pre-intervention phase, but it improved significantly in the post- intervention phase, this applied to all related areas of knowledge. On the same line **Khalaf et al., (2017)**, entitled (Effect of Training Program on Nurses' Competent Practices Towards Children Receiving Blood Transfusion) who found that nurses' knowledge regarding proper methods of blood and its components transfusion are deficits of all aspects of blood transfusion .This may be due to lack of training and information of nurses about the impact of the disease and in response to this problem,

The present study has also revealed that nurses` knowledge about management was unsatisfactory in the pre-intervention phase, but it has improved significantly in the post- intervention phase. This applied to all related areas of knowledge as ,intensive wound care, correcting the coagulopathy with PC replacement ,parenteral nutrition (PN) with protein and calories as tolerated ,supportive care for the ophthalmic functions and follow up ,prevention of infection and follow up and parental education. These results are in agreement with **Martí-Carvajaletal (2012),**entitled **(**Human recombinant protein C for severe sepsis and septic shock in adult and pediatric patients),who have emphasized that the Protein C (PC) deficiency is a rare but life-threatening bleeding disorder that can present in the children .Furthermore **Kabinda et al., (2014)** reported that training and education are essential for all nurses caring for children with Protein C deficiency therapy enhanced continuity of care and as well promoted better child outcome and reduced child and family members` anxiety levels.

On the other hand, the current study assessed nurses' practice for infants` with Protein C (PC) deficiency. The study findings demonstrated very low levels of practice among them before implementation of the program. This was particularly evident in crucial tasks such as preparation skills prior intensive wound care, correcting the coagulopathy, pain management and supportive care for the ophthalmic issue. Overall, none of the nurses had incompetent total practice at the pre- intervention phase. The minority of nurses had competent total practice at pre- intervention. This deficient practice revealed among the present study nurses before intervention is certainly linked to the previously mentioned low level of satisfactory knowledge among them. Both together, deficient knowledge and practice would certainly have a negative impact on the quality of nursing care provided by the studied nurses. As emphasized by **Khalaf et al., (2017)**, who have also mentioned that the poor blood transfusion practice is likely to play a role in the morbidity and mortality of patients who receive blood transfusion. So that, nurses with specialist knowledge, practices and expertise in pediatric hematology (Protein C deficiency) are needed in position where they are able to develop nursing practice.

Overall, the present study revealed that the intervention study about protein C deficiency was effective in achieving better knowledge and practice among studied nurses. This effect was further confirmed by multivariate analysis, which has revealed that intervention study attendance was the strongest statistically significant positive independent predictor of nurses' knowledge and practice scores at the intervention. These findings was supported by **Khalil et al., (2013)** who revealed in their study that continued nursing education programs for nurses increase their knowledge and practice where there was an improvement in nurses' practice after the attendance continuing nursing education sessions.

However, the retention of knowledge and practice in the current study was low normally because protein C deficiency remains a rarely diagnosed. Therefore, the intervention study was successful in improving nurses' knowledge and practice, which could be attributed to the structure, content and process of the intervention. As highlighted by **British Committee for Standards in Haematology (2012)** the primary responsibility of the nurse educator is to build quality program on the existing knowledge and practice and to translate their teaching needs into systematic learning experience. Learning in nursing must be relived to the goal and objectives of the program.

The intervention study for nurses who caring for infants with protein C deficiency (PCD) can provide them with a higher level of knowledge and practices and would improve the ability of the nurses in health decision-making and consequently would cause changes in their behavior .

5. CONCLUSION

Based on results of the present study, after intervention study, nurses had satisfactory knowledge and competent practice, than before intervention, regarding protein C deficiency for infants. As well as, there was a statistically significant relation between nurses' knowledge and practices scores with their age, qualifications and years of experience pre and post intervention.

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 nurses' knowledge, as well as reinforce proper practices related to protein C deficiency in pediatric units.

-Continuous supervision and evaluation for nurses is needed to determine any defect related to knowledge or practices, Policies and strategies for hospital related to protein C deficiency must be setting.

Future studies;

Severe protein C deficiency remains a rarely diagnosed condition, an emphasis on nurses` experience and management would enhance the current state of the science and provide a much-needed window into interventions aimed at improving the life of children with protein C deficiency.

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