Instructional Module for Nurses regarding Care of Children with Hemophilia

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Abstract

Hemophilia is a coagulation disorder arising from a genetic defect of the X chromosome; the defect can either be inherited or result from spontaneous gene mutation. In each type of hemophilia (A, B, C, and willebrands disease), a critical coagulation protein is missing, causing children to bleed for long periods of time before clotting occurs. **Aim:** this study was a quasi-experimental aimed to evaluate the outcomes of the instructional module on the studied sample. Setting: the study was carried out at hematology departments at children's hospitals affiliated to Ain Shams University Hospital (Al Demerdash) Cairo University Hospital (Abo Elreesh) and Benha children's Hospital affiliated to ministry of health. **The Sampling**: A convenient sample involved Fifty nurses from the previously mentioned settings. **Tools of data collection:** involved a structure questionnaire to assess nurses' knowledge and observational checklists to assess performance regarding care of children with hemophilia and instructional module about hemophilia. **Results:** Slightly less than half (46.0% &48%) of the studied nurses their ages were 30 years and more, had nursing diploma from technical institute of nursing respectively, less than two thirds of the nurses (62%) had experience ≥ 6years, Post implementation of the instructional module most of the studied nurses had satisfactory level of knowledge and competent practice in comparison to pre implementation of the instructional module. **Conclusion:** The designed instructional module led to significant improvement of knowledge and performance of the studied nurses towards care of hemophilic children**. Recommendations:** continuous refreshing training should be provided for all of nurses working in hematological departments and caring of children with hemophilia. A manual guided booklet about hemophilia should be available to all nurses working in hematological departments.

Key words: Hemophilia, instructional module, children, nursing care.

Introduction

Hemophilia (he-mo-FIL-e-ah) from the Greek haima αἷμα 'blood' and philia φιλία 'love' is a group of hereditary genetic disorders that impair the body's ability to control blood clotting or coagulation, which is used to stop bleeding when a blood vessel is broken. It also is an inherited condition that is present at birth and usually occurs mostly in boys (***Kaufman et al., 2013***).

Normally, blood cells and substances leading to protect and stop the child's body from bleeding. These include platelets, clotting factors, vitamin K and fibrinogen form the clot. Clotting factors are proteins made by cells that line the blood vessel walls. Platelets are a type of blood cell made in the bone marrow that helps blood to form a blood clot ***(Friedman et al., 2013)***. When a blood vessel is injured, the blood cells and substances stick to the cut area and form a clot. The clot acts as a plug to stop the bleeding and covers the blood vessel injury while it heals. Blood clots normally form

after a blood vessel injury, such as a cut or during a nosebleed (*Chitlur and Kulkarni, 2011*).

According to the World Federation of Hemophilia *(WFH,2012)* the annual global surveys indicate that the number of patient with hemophilia in the world is approximately 400,000 Hemophilia incidence worldwide is estimated to occur in 1:7,500 live male births; all races and ethnic groups are affected. Factor VIII (8) deficiency is four times more common than factor IX (9) deficiency but the clinical presentations and inheritance patterns are the same *(Stonebreaker et al., 2012)*. Currently, about 20,000 patient in the United States have hemophilia, about 70% of hemophilia patients have less than one percent of the normal amount of clotting factors and thus, have severe hemophilia. Hemophilia affects mostly boys, about 1 in every 5,000-10,000 is born with it. Girls are more rarely affected. A male can't pass the gene for hemophilia to the sons, though all the daughters will be carriers of the disease gene. Each male child of a female carrier has a 50% chance of having hemophilia *(Stonebraker et al., 2012)*. The two major forms of hemophilia (A, B) occur much more commonly in males than in females. Hemophilia A is the most common type of the condition; 1 in 5,000 males worldwide are born with this disorder. Hemophilia B occurs in approximately 1 in 20,000 newborn males' worldwide *(Ghai et al., 2010)*.

Children with Hemophilia in Egypt an estimated 20,000 child live with bleeding disorders. About 1,500 are registered with six hemophilia treatment centers which located mainly in major cities. The Egyptian government provides approximately 38% of healthcare costs *(Said, 2013)*. Most hemophilia children are treated with plasma or cryoprecipitate, resulting in a high risk of transfusing blood-borne diseases like HIV and hepatitis B and C. Factor concentrate is available only in limited quantities, and patients children must travel long distances to get it *(Srivastava et al., 2013)*.

Though there is no cure for hemophilia, it can be controlled with regular infusions of the deficient clotting factor, i.e. factor VIII in hemophilia A or factor IX in hemophilia B. Factor replacement can be either isolated from human blood serum, recombinant, or a combination of the two. Some hemophilic children develop antibodies (inhibitors) against the replacement factors given to them, so the amount of the factor has to be increased or non-human replacement products must be given, such as porcine factor VIII (*Baker et al., 2013*).

Most nurses have little experience with them and little knowledge of how to care for children who have hemophilia. Children with hemophilia and their families who are knowledgeable about their disorder quickly notice caregivers' lack of knowledge and it makes them uncomfortable (*Leticia et al.,2012*). Learning about bleeding disorders can vastly improve the care that is provided and increase children' confidence in the care, so that the current study will be carried out in an attempt to promoting the children's heath status through helping the nurses to increase their working knowledge and to change their pitfalls practices related to hemophilic children through initiation of an instructional module *(Khair, 2013*).

Aim of the study:

The current study was carried out to evaluate the outcomes of an instructional module for nurses caring for children with hemophilia.

Hypothesis:

Instructional module has a positive effect on nurses' knowledge and performance regarding health care of children with hemophilia.

Subjects and methods:

Technical Design:

Research design:

A quasi- experimental design was utilized to conduct the instructional module

Research Setting:

This study was conducted at Hematology Departments in Children's Hospitals affiliated to Ain Shams University, Cairo University and Benha Children Hospital affiliated to Ministry of Health.

Research sampling:

Convenient sample was included (50) nurses were working in hematology departments in pediatric hospitals at the previously mentioned study settings**.**

Tools of Data Collection:

Data will be collected through the following tools:

I - Structured Questionnaire Form:

It was prepared and designed by the researcher after reviewing the recent related literature. It was written in Arabic language and contents validated by supervisors. This tool was divided into two parts:

Part 1:

This part was concerned with socio-demographic characteristics of nurses as regards: age, marital status, educational level, years of experience and residence.

Part 2:

This part was concerned with nurses' knowledge and contained 10 items about hemophilia such as definitions, types, causes, pathophysiology, clinical manifestations, diagnosis, treatment, complication, prevention and nursing care of hemophilia**.**

Scoring system of the studies sample knowledge:

For the 10 knowledge items, a correct response was scored one and the incorrect zero for each item knowledge, these scores were converted into a percentage score, the subject knowledge was considered

- Satisfactory $\geq$ 85 %

- Unsatisfactory $<$ 85 %

II- *Observational checklists:*

It was constructed through standardized checklists applied in order to assess the nurses' performance regarding care of children with hemophilia (pre and post instructional module implementation). Observational checklists covered 7 items, these were the following procedures, insertion of I.V catheter, blood sample aspiration, blood transfusion, oral and skin care measures to avoid bleeding and infection.

***Scoring System*:**

For all 7 procedure items, each procedure was scored according to key answer to weighing of each step that makes a total score (equal 100%). Each right step was getting one score while wrong step or not done was getting zero score. The scoring system of nurses' practice was classified into competent $\geq$ 85% and incompetent $<$ 85%.

III - *Instructional Module:*

The Instructional module contained 7 sections. These sections explained definitions of hemophilia, types, causes pathophysiology, clinical manifestations, diagnosis, medical treatment, complications, and nursing intervention for children with hemophilia. After each section there was

questions to enable the nurses' self evaluation. Pre and post tests was done for nurses, it contained all types of questions (define, complete, true or false and matching. Each question had a written score and defined time written upon it.

Scoring system:

Each nurse should successfully complete and pass post test for this module with a score of ≥ 85 % towards knowledge about hemophilia.

The operational designs:

A) *Preparatory phase:*

During this phase, the researcher reviewed current local and international related literature, which helped to be more acquainted with the topic, and with the process of tools' construction.

Pilot study:

A pilot study was included 10 % of the total study subjects (5 nurses) then they were excluded from the study sample. Results of data obtained from the pilot study helped to modify the tools where some items added such as daily activities of the child. The final form of the tools was then obtained and the time needed for completing each tool was also determined.

B) Implementing Phase:

Field of Work:

The actual field work was carried out over 6 month's period from the first of Mars 2014 up to the end of August 2014. The investigator was available daily from 9.00 AM to 1.30 PM, for 3days / week by rotation of study settings.

Module implementation phase:

The instructional module designed for this study has been implemented throughout 7 sections for the fulfillment of the expected objectives theoretical and practical. Each section took 60-90 minute. These sections explained definition of hemophilia, types, causes pathophysiology, clinical manifestations, diagnosis, medical treatment, complications, and nursing intervention for children with hemophilia and Performance checklists such as blood transfusion checklist. After each section there was questions to enable the nurses' self evaluation. Pre and post tests was done for nurses, it contained all types of questions (define, complete, true or false, matching). Each question had a written score and defined time written upon it.

C) Evaluation phase :

The evaluation of the instructional module was done by using a pre/ post-test format before the researcher implement the instructional module and after finishing the all module, as well as immediately after each section of the all module, each section had evaluation to assess feedback of the nurses' knowledge & practice concerned children with hemophilia.

Administrative design:

Official letters clarifying the title, aim and expected outcomes of the study was obtained from the Dean of the Faculty of nursing, Ain Shams University. It was addressed to each director of the previously mentioned hospitals to obtain their approval to conduct the study.

Ethical Considerations:

Approval was taken from the director of each hospital. Then, the oral consent was taken from each nurse to participate in the study after explaining the objectives of the study. Nurses were given an opportunity to

refute to participate and they were notified that could withdraw at any stage of the research, also they were assured that all information would remain confidential, and used for the research purpose only

Statistical design:

The collected data were revised, tabulated and analyzed using the number and percentage. Statistical analysis was done using SPSS (Statistical Package for Social Science) program versions 20. Data were presented using descriptive statistics in the form of frequencies and percentages for qualitative variables, means and standard deviations for quantitative variables, qualitative variables were compared using Chi-Square test for relation. When $p < 0.05$ there is a statistical significance and when $p < 0.001$ there is highly statistical significance $p > 0.05$ there is no statistical significance.

Result:

Table (1): Distribution of the Studied Nurses according to their knowledge about identification of Hemophilia pre /post instructional module implementation (n=50).

Knowledge items	Pre				Post				Chi-square test	
	Satisfactory		Unsatisfactory		Satisfactory		Unsatisfactory		X^2	p-value
	No.	%	No.	%	No.	%	No.	%		
Definition of hemophilia.	31	62.0	19	38.0	46	92.0	4	8.0	12.705	**<0.001
Types of hemophilia.	11	22.0	39	78.0	46	92.0	4	8.0	49.980	**<0.001
Causes of hemophilia.	3	6.0	47	94.0	49	98.0	1	2.0	84.776	<0.001
Pathophysiology of hemophilia.	23	46.0	27	54.0	47	94.0	3	6.0	27.429	**<0.001
Clinical manifestations of hemophilia.	8	16.0	42	84.0	48	96.0	2	4.0	64.935	**<0.001
Diagnosis of Hemophilia.	3	6.0	47	94.0	44	88.0	6	12.0	67.483	**<0.001
Methods of treatment of hemophilia.	18	36.0	32	64.0	47	94.0	3	6.0	36.967	<0.001
Complications of hemophilia.	20	40.0	30	60.0	47	94.0	3	6.0	32.972	**<0.001
Prevention of hemophilia.	10	20.0	40	80.0	47	94.0	3	6.0	55.855	**<0.001

** Statistical significant $p \leq 0.001$

Table (2): Distribution of the studied nurses' knowledge regarding nursing intervention of skin problems and infection pre/ post implementation of the instructional module (n=50).

Nursing intervention for skin problems & infection.	Pre				Post				Chi-square test	
	Satisfactory		Unsatisfactory		Satisfactory		Unsatisfactory		X^2	p-value
	No.	%	No.	%	No.	%	No.	%		
Nursing intervention to avoid inflammation of the skin.	11	22.0	39	78.0	47	94.0	3	6.0	53.202	**<0.001**
Nursing intervention in the occurrence of skin inflammation.	15	30.0	35	70.0	46	92.0	4	8.0	40.395	**<0.001**
Measures to avoid infection of children with hemophilia.	11	22.0	39	78.0	48	96.0	2	4.0	56.594	**<0.001**
Nursing intervention in the occurrence of infection in children with hemophilia.	14	28.0	36	72.0	44	88.0	6	12.0	34.524	**<0.001**

Statistical significant $p \leq 0.001$

Table (3): Distribution of the studied nurses according to their performance related to blood transfusion pre / post implementation of the instructional module (n=50).

performance items	Pre				Post				Chi-square test	
	Competent		Incompetent		Competent		Incompetent		X^2	p-value
	No.	%	No.	%	No.	%	No.	%		
Cheek vital signs	22	44.0	28	56.0	48	96.0	2	4.0	32.190	**<0.001**
Wash hands and wear gloves	15	30.0	35	70.0	48	96.0	2	4.0	46.718	**<0.001**
open blood tube to evacuate the air and blood come to the end	21	42.0	29	58.0	48	96.0	2	4.0	34.081	**<0.001**
Connect the blood tube into the child transfusion Rate to 10 drops /Min in the first 15 minutes	25	50.0	25	50.0	47	94.0	3	6.0	24.008	**<0.001**
cheek vital signs	48	96.0	2	4.0	49	98.0	1	2.0	0.344	0.558
Increase blood transfusion rate if no reactions	29	58.0	21	42.0	48	96.0	2	4.0	20.384	**<0.001**
Remove gloves	32	64.0	18	36.0	44	88.0	6	12.0	6.634	**0.010**
Wash hands	26	52.0	24	48.0	43	86.0	7	14.0	11.968	**<0.001**
Record	24	48.0	26	52.0	45	90.0	5	10.0	18.700	**<0.001**
Check vital signs	34	68.0	16	32.0	40	80.0	10	20.0	1.299	**0.254**

Statistical significant $p \leq 0.001$

Table (4): Distribution of the studied nurses according to their practice related to safety measures to prevent bleeding pre / post implementation of the instructional module (n=50).

performance items	Pre				Post				Chi-square test	
	Competent		Incompetent		Competent		Incompetent		X^2	p-value
	No.	%	No.	%	No.	%	No.	%		
Instruct the child to avoid aggressive playing such as football and jumping.	34	68.0	16	32.0	47	94.0	3	6.0	10.981	**0.001**
avoid giving the child hard food to protect him from gum bleeding.	28	56.0	22	44.0	47	94.0	3	6.0	19.253	**<0.001**
wash the mouths of the child with water and bicarbonate solution and apply lubricant to lips to prevent cracking and occurrence of bleeding.	16	32.0	34	68.0	47	94.0	3	6.0	41.227	**<0.001**
Never using of aspirin.	26	52.0	24	48.0	50	100.0	0	0.0	31.579	**<0.001**
proper pressure over the site of blood sample aspiration.	34	68.0	16	32.0	48	96.0	2	4.0	13.279	**<0.001**
Avoid the hard cough and consult physician for the proper antitussive.	29	58.0	21	42.0	44	88.0	6	12.0	11.416	**0.001**
giving the proper clotting factor before tooth extraction or any surgical operation.	48	96.0	2	4.0	50	100.0	0	0.0	2.041	0.153
avoid intra- muscular injection	50	100.0	0	0.0	50	100.0	0	0.0	-	-
Children from 2-5 years can protect them from contusion and wounds by using shoulder and knee bandages.	6	12.0	44	88.0	39	78.0	11	22.0	44.000	**<0.001**
Remove all furniture that have sharp edges, and all sharp equipment	17	34.0	33	66.0	36	72.0	14	28.0	14.492	**<0.001**

Statistical significant $p \leq 0.001$

Table (5): Correlation between total knowledge score and total practice score (n=50).

Total performance score	Total knowledge score		
		R	P
	Pre training	0.375	<0.05*
	Post training	0.861	<0.001**

Statistical significant $p \leq 0.001$.

As observed from **table (1)** that, more than three fifths (62 %)of the studied nurses had satisfactory knowledge related to definition hemophilia pre-module implementation, in contrast this percentage increased up to 92% post instructional module implementation. Moreover, the cause of hemophilia was unknown for 94 %of the nurses pre- instructional module implementation, vice versa it was known for majority of them (98%) post instructional module implementation. There were statistically significant difference pre/post instructional module implementation with P value < 0.001.

Table (2) clarified that, there was an improvement of the studied nurses' knowledge to avoid inflammation of the skin and infection for children with hemophilia as most (94% & 96%) of them had satisfactory knowledge post implementation of the instructional module respectively compared to pre implementation of the module the percentages were 22% & 28% respectively .This table reflected statistical difference as P-value < 0.001.

Discussion:

Hemophilia is a rare disorder that is complex to diagnose and to manage. Children with hemophilia are at higher risk bleeding and other health hazards than healthy children. In caring of children with hemophilia, nurses are key components of the health care team, who have excellent relationships with children, and their families

Table (3) revealed that the most (96%) of the studied nurses had competent practices related to wash hands and wear gloves for blood transfusion procedure post implementation of the instructional module compared with pre implementation of the instructional module the percentage was 30 % of them in relation to the previous item. This improvement proved statistical significant as (p<0.001).

Table (4) clarified that, slightly less than one third (32%) of the studied nurses had incompetent practice related to proper pressure over the site of blood sample aspiration pre implementation of the instructional module, but post implementation of the instructional module most (96%) of nurses had competent practices in relation to the previous item. This improvement proved to be statistically significant (p < 0.001).

Table (5) showed that, there was a statistical significant differences pre and post instructional module implementation between the nurses' knowledge and their practices regarding care of children with hemophilia where p-value <0.001.

evaluating health care outcomes and quality of life impacts on new treatment options (*Colvin et al., 2013*).

The present study, showed that the studied nurses had an improvement of their knowledge related to identification of hemophilia, post implementation of the module as more than three fifths of the studied nurses had satisfactory knowledge related to definition hemophilia pre-module

implementation, in contrast this percentage increased post module implementation. Moreover, the causes of hemophilia were unknown for most of nurses pre-module implementation, vice versa it were known for most of them post module implementation. This result was in accordance with **Saad, (2012)** who reported that most of nurses had satisfactory knowledge regarding definition and causes of hemophilia compared with all of them after application of nursing management of children with hemophilia according to basic standard.

The instructional module was successful in upgrading knowledge of nurses regarding nursing intervention to avoid inflammation and infection of the skin for children with hemophilia, the findings revealed that, more than three quarters of studied nurses had unsatisfactory knowledge pre instructional module application, on the other hand most of the nurses had satisfactory knowledge post instructional module application. The present study findings agreed with the results reported by **Rekha, (2008)** entitled knowledge, attitude & practice of first aid among women in rural area, who stated that*,* the responsibility of the caregivers for the child care to make sure that dangerous substances are out of reach of children and protect them from inflammation and infection of the skin .

Regarding nurses practice related to blood transfusion, the present study revealed that the most of the studied nurses had competent performance related to wash hands and wear gloves for blood transfusion procedure post implementation of the instructional module, compared to less than one third of them pre implementation of the module. This improvement proved statistical significant as (p<0.001). The researcher opinion confirmed the importance of safety measures for blood transfusion and infection control especially for children with hemophilia and protect them from blood - born infection. The researcher opinion was

supported by **Hamid,** (**2004**) who mentioned that, transfusion of blood products derived from multiple units of pooled blood can transmit HIV and accounted for the initially high prevalence of HIV infection among patient with hemophilia.

Regarding safety measures to prevent bleeding for children with hemophilia, the present study assessed the nurses performance and clarified that slightly less than one third of the studied nurses had incompetent practice related to proper pressure over the site of blood sample aspiration pre implementation of the instructional module, but post implementation of the instructional module most of them had competent practices in relation to the previous item. These results were in accordance with **Saad, (2012)** who reported that slightly more than two thirds of the nurses were competent for obtaining blood sample aspiration before standard application but after the standard all nurses were competent. In the same line **Hamid, (2004)** mentioned that, as infants begin to become mobile, all items around must be safe. Also toddlers fall a lot and they should be observed for any pain or discomfort, which may indicate bleeding in a joint or a muscle. A critical part of every child development is playing, but contact sports such as football are strongly discouraged, socialization and active participation should be encouraged. Early psychological intervention helps families to achieve a balance between over protection and permissiveness.

As regards (the pre and post instructional module application) between the nurses' knowledge and their performance regarding care of children with hemophilia, there was a statistical significant where p <0.001. The researcher certified further courses for nurses and continuous training to ensure the best care for children with hemophilia.

Conclusion:

The results revealed significant improvement of knowledge and practice of studied nurses regarding care for children with hemophilia post the instructional module application, and these findings reflect that, the instructional module has a module has a positive effect on nurses' knowledge and practice.

positive effect on nurses' knowledge and practice.

Recommendations:

Based on results of this study, the following recommendations can be suggested:

1. Instructional module about hemophilia should be provided to all newly employed nurses in hematology departments.

2. Periodic continuous training should be provided for the nurses working in hematology departments and better co-operation of health professionals to provide continuous training about first aid emergency situations management and bleeding of children with hemophilia.

3. Expand, empower and update services in hematology departments and outpatient clinics to include prevention, control and management of hereditary blood diseases specially hemophilia.

4. Adopting a national strategy aimed to promote health status of children with hemophilia, prevent and minimize their exposure to blood born infection specially hepatitis C, or B and HIV.

References

Baker, J.R., Riske, B., Drake, J.H. (2013): US Hemophilia Treatment Center population trends 1990-2010: patient diagnoses, demographics, health services utilization, Hemophilia. 19 (1); 21- 6.

Chitlur, M. and Kulkarni, R. (2011): Hemophilia and related bleeding disorders. In ET Bope, et al., eds., Conn's Current Therapy 2011, Philadelphia, Saunders pp. 428-434.

Colvin, B . Astermark, J. Fischer, K. Gringeri, A . Lassila, R. Schramm, W. Thomas, A. and Ingerslev, J. (2013) : inter disciplinary working group. European principles of hemophilia care. Hemophilia 14(2):361-7411.

Friedman, M. White, B. and Dougall, A. (2013): An audit of the protocol for the management of patients with hereditary bleeding disorders undergoing dental treatment. J Disab Oral Health; 10 (4):151-55.

Ghai,O. paul, V. and Bagga, (2010): Essential Pediatrics, 7th ed, Thomson press, India, p-78.

Hamid, H. (2004): A thesis Submitted in partial fulfillment for the requirement of clinical MD degree in pediatrics and child health, University of Khartoum Graduate College Medical &health studies board, 15-87.

Kaufman, R. Fay, P. Popolo, L. Ortel, T. Marder, V. Aird, W. Bennett, J. Schulman, S. and White, G. (2013): Factor V and factor VIII, 6th ed Hemostasis and Thrombosis: Basic Principles and Clinical Practice, PA Lippincott Williams and Wilkins; Philadelphia, 179-96.

Khair, K. (2013): An exploration of the lived experience of boys with hemophilia, doctorate thesis, 20.

Leticia Riley, Mack Womack, and Susan Zappa, (2012): Emergency room care" national hemophilia foundation, nursing working group nurses' guide to bleeding disorders, N. Engl. J. Med. 357 (6): 603–60.

Rekha, F. (2008): Knowledge, Attitude & Practice of First Aid Among Women in Rural Area, Master Thesis, Community Medicine, University Of Health Sciences Bangalor, Karnataka, India; Pp.160.

Saad, A (2012): Nursing management of children with hemophilia according to basic standard, doctorate thesis, 3(2):168 -187.

Said, I . (2013): Evaluation of the oral and dental health of children with hemophilia in JordanThesis submitted for the degree of M.Sc. in Dentistry / Pediatric Dentistry,Jordan University of Science and Technology,pp.14-67.

Srivastava, A.; Brewer, A. and Mauser-Bunschoten, E (2013): Guidelines for the management of hemophilia. Hemophilia; 19:e1-e 47.

Stonebraker, J. Bolton-Maggs , P. Soucie, J. Walker, I. Brooker, M . (2012) : A study of variations in the reported hemophilia A prevalence around the world, Hemophilia; 18 (3): e 9 20-32.

World Federation of Hemophilia, (2012): Guidelines for the management of hemophilia 2nd,Blackwell Publishing in Hemophilia; Epub 6 JUL, 7-69.