Effect of Implementing an Educational program Regarding Vacuoles, E1-enzyme, X-linked, Auto-inflammatory and Somatic Syndrome on Nurses' Knowledge in Intensive Care Units

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Abstract

Background: Vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic (VEXAS) syndrome is recently identified systemic autoinflammatory disease that manifests in adults and has a high morbidity and mortality rate. **Aim**: to evaluate the effect of implementing an educational program regarding vacuoles, E1-enzyme, X-linked, auto-inflammatory and somatic syndrome on nurses' knowledge in intensive care units. **Design**: quasi-experimental research design was utilized. **Setting**: The study was accomplished at the Sohag Main University Hospital, in Critical Care and General ICU. **Subjects**: Convenient samples of 70 nurses were recruited from general and critical care ICUs **Tool I:** Structured Interview Questionnaire Assessment Included Nurses' Demographic Data. **Tool II:** Nurses' Knowledge Pre-Post Educational Program Assessment. **Results**: the mean knowledge score was 7.31±4.1 in the pre-test versus 18 ±1.7 in the post test with high statistically significant difference and about 6% of nurses had satisfactory knowledge level in pretest versus 84% of them after implementation of educational program. **Conclusion:** After the educational program was put into place, the majority of nurses in the study group had a good level of knowledge, and their overall mean scores significantly improved **Recommendation:** The current study should be replicated on larger probability samples.

Key words: Auto-Inflammatory, Educational Program, E1-Enzyme, Intensive Care Units, Nurses' Knowledge, Somatic Syndrome, Vacuoles, X-Linked.

Introduction

A rare condition called vacuoles, E1-enzyme, X-linked, auto-inflammatory, and somatic syndrome (VEXAS) syndrome results in hematologic (blood) and inflammatory symptoms. The disease is acquired later in life and is brought on by mutations in the blood cell Ubiquitin-like Modifier Activating Enzyme 1 (UBA1) gene. Patients do not transmit the illness to their offspring. It was first discovered in 2020 and primarily affects men over 50 years **(Stephanie, 2020).**

Innate immune system dysregulation is the cause of autoinflammatory syndromes (AIS). Over the past thirty years, more than thirty monogenic AIS have been discovered and described thanks to the growth and greater use of genetic testing. Patients who have clinical signs of AIS but do not have genetic abnormalities are more difficult to treat. As a result, non-germline somatic mutations have been investigated as a possible cause of these traits, especially in people with later-onset illness symptoms and family members who are unaffected **(Koster et al., 2024).**

Recurrent fever, weight loss, skin lesions, and other inflammatory presentations in several organ systems are typical clinical symptoms. Due to the overlap of symptoms with other autoimmune and inflammatory disorders, diagnosing VEXAS syndrome can be difficult and frequently necessitates a thorough medical evaluation before a conclusive diagnosis is reached **(Najdi & Karaa, 2024).**

Familial Mediterranean fever, and tumor necrosis factor receptor–associated periodic syndrome, and adenosine deaminase deficiency are the most prevalent genetic causes of inflammation. UBA1 variations are acquired and develop later in life, in contrast to previously identified genetic causes of inflammation. Because UBA1 is X-linked, men and women with X chromosomal monosomy are more likely to have phenotypic effects. The molecular diagnosis is more important in determining the prognosis of VEXAS syndrome than the clinical diagnosis. Despite the fact that a genotype-driven approach was used to identify the first three VEXAS syndrome patients, subsequent

ascertainment has been phenotype-driven **(David et al., 2023).**

Since nurses are the ones who constantly care for inpatients, they can be the most trustworthy individuals to disseminate their knowledge and practices for the health and welfare of the populace, humanity, or country. The goal of the nursing profession has always been to improve people's "health, relieve their pain and misery, advocate for the weak and the susceptible, and educate the people to attain a better quality of life **(Jaleta et al., 2021).**

Significance of the Study

Although the prevalence of VEXAS syndrome is unknown, it is probably underreported. At first, only males with somatic mutations in UBA11 were reported to have VEXAS syndrome. However, due to acquired somatic mutations and X monosomy in hematological cells, we and others reported VEXAS syndrome in female patients. The recruitment and buildup of inflammatory cells in this tissue is supported by clinical symptoms such as fever, neutrophilic skin lesions, arthralgias, pulmonary inflammation, chondritis, and vasculitis **(Kosmider et al., 2024).**

According to a 2023 study, about 1 in 13,591 adults are affected. The illness is more prevalent in men over 50, according to an analysis of health information from over 160,000 persons. VEXAS was found in 1 in 26,238 women and 1 in 4269 males over the age of 50, according to earlier estimates of prevalence **(Najdi & Karaa, 2024)**.

This study is the first study to implement an educational program about VEXAS syndrome for the ICU nurses; it could be beneficial in many ways. First, it will provide a database that can be utilized by health team members to raise staff awareness about VEXAS syndrome. Second, health professionals can be able to identify and deal with VEXAS syndrome manifestations. It is also hoped that this effort will generate attention and motivation for further research into this area.

Aim of the Study

The Aim of the current study was to evaluate the effect of implementing educational program regarding vacuoles, E1-enzyme, X-linked, auto-inflammatory and somatic syndrome on nurses' knowledge in intensive care

Research Hypothesis

To fulfill this, aim the following research hypothesis was formulated:

- Nurses' knowledge regarding vacuoles, E1-enzyme, X-linked, auto-inflammatory, and somatic syndrome after education is expected to be higher than that before education.

Subjects and Method

Design

Quasi-experimental research (pre-posttest) research design was used to conduct this study. This design was used to assess nurses' knowledge regarding vacuoles, E1-enzyme, X-linked, auto-inflammatory and somatic syndrome in intensive care units

Setting

The study was conducted in General and Critical Care ICUs at Sohag Main University Hospital.

- General ICU: it has 33 nurses (nursing institute) and 8 head nurses (bacholar degree)
- Critical Care ICU: it has 22 nurse (nursing institute) and 7 head nurses (bacholar degree)

Subjects

- A convenient sample included all available critical care nurses who work in the previous setting, the total number of nurses was 70 nurse

Inclusion Criteria

-All Critical care nurses who were directly involved with patient care in General and Critical Care ICUs at Sohag Main University Hospital.

Study Tools

Two tools were used in this study after reviewing the related national and international literatures.

Tool I: Structured Interview Questionnaire : (Khitri et al., 2024).

This tool was developed by the researcher after reviewing the related literature. It is composed of three parts. It was used to assess personal and socio-demographic data of nurse, and to assess nurses' level of knowledge regarding caring for patients. It was written in an Arabic language to suit all the educational levels of the studied nurses. It was filled by the studied nurses themselves. It included two parts:

- Part I: Personal characteristics which include age and sex
- Part II: Socio-demographic data which include: level of education, years of experience, working unit, marital status, and previous training courses

Tool II: Nurses' Knowledge Pre-Post Educational Program Assessment: (**Roy,2023**),& (**Sterling,2023**)

It was used to assess nurses' knowledge regarding VEXAS syndrome, it consists of definition of VEXAS syndrome, causes, manifestations of VEXAS syndrome, treatment of VEXAS syndrome, complications are associated with VEXAS syndrome, laboratory tests and imaging studies are used, how to differentiate VEXAS syndrome from other rheumatologic disorders, primary nursing intervention to manage VEXAS syndrome.

Scoring System:

Correct answers receive one mark, while incorrect answers receive zero. The following categories will be applied to the scoring system's results:

- Satisfactory knowledge level: Equal to or more than 70%
- Unsatisfactory knowledge level: less than70%

Methods

There were three primary phases of the investigation.

l- Preparatory Phase:

After explaining the purpose and scope of the study, permission to perform it was granted by the dean of Sohag University's nursing faculty as well as hospital accountable authorities, including the head of the anesthesia department, who serves as the principal of all intensive care units. Development of instruments following an evaluation of relevant research. Five jurors, comprising two medical professionals and three critical care nurses from Sohag University, evaluated the tools for face and content validity. The validity index was 0.88, and nothing had changed. There were no reported changes. The Cronbach's Alpha test was used to assess the tools' reliability. With a dependability score of 0.98, the results demonstrated the instruments' stability and consistency.

Ethical Considerations

The nursing faculty's ethics committee authorized the research idea, (**the ethical code is 295)**. Study participants were not at risk when the research was being applied. The study adhered to standard clinical research ethics. After outlining the nature and goals of the study, the nurse who agreed to participate gave their informed consent. Anonymity and confidentiality were guaranteed. Participants in the study were free to decline participation or to leave the study at any time. The privacy of study nurses was taken into account when gathering data.

To verify the tool's viability and applicability, **a pilot study** was carried out on seven nurses who satisfied the predetermined selection criteria; no necessary modifications were made; the seven patients in the pilot study were not included in the study.

II-Implementation Phase:

The Researcher introduced herself to nursing staff in each unit and explained the research points. After nurses agreed to participate in the research, the researcher asked nurses to take an appointment to arrange meeting with each group of them; for example, after ending their shift to start the first session of the educational program**.**

First session: The participants filled out the structured interview Questionnaire sheet; it was checked by the nurses in this session without giving them any information about VEXAS syndrome. This session took about half an hour. At the end of session, the researcher took an appointment for the second session.

The second session:

The researcher gave an introduction about the syndrome and then explained everything related to VEXAS syndrome, such as the definition, causes, manifestations of VEXAS syndrome, treatment, complications associated with VEXAS syndrome, laboratory tests and imaging studies, how to differentiate VEXAS syndrome from other rheumatologic disorders, and primary nursing intervention to manage VEXAS syndrome. During this session the researcher uses teaching methods: videos, real-life case studies, and interactive questions and answers. This session took about an hour and was performed on the same day as the first session. The researcher at the end of the session asked the participants for a new appointment to the final session.

The third session: it was performed after three days of the previous session. In this session feedback was taken from nurses by writing down in Post test-sheet (Part I II). This session took about 30minutes

III-Evaluation Phase:

The participants were evaluated twice, the first time before providing them with education and the second time after education and feedback was documented in the questionnaire.

Statistical Analysis

To make data appropriate for computer processing, it will be coded and converted into a properly created format. The software program SPSS, version 20, will be used for statistical analysis. The acquired results were summarized as frequency using mean ± SD, number, and percentage, and the statistical significance between two variables was determined using the t-test. To ascertain the significance between

variables, use the chi-square test. To assess the significance of change over time, use the paired T-test.

Limitation of the Study

Due to the small sample size and lack of demographic representation, the study's findings could not be broadly applied. The study may only assess the immediate knowledge changes without long term follow up to evaluate the sustained impact especially because the studied syndrome is newly identified.

Results

Table (1): Reveals that 54.3% of the studied nurses were females. In relation to age, 65.7% are in the age group of 18-35 years old. As regards the level of education, 60% of them had bachelor's degrees, 14.3% had master's degrees in nursing, and 2.9% of them had doctoral degrees in nursing. Concerning marital status, about 54.3% were single, and 42.9% were married. Regarding the years of experience, 58.6% of them worked for 3-10 years in the nursing field**.**

Table (2): Shows that there are highly statistically significant differences in nurses' knowledge mean scores between pre- and post-implementation of the teaching program regarding VEXAS syndrome.

Figure (1): Illustrates that about 6% of nurses had satisfactory knowledge in the pretest versus 84% of them after the implementation of the educational program, with a high statistically significant difference, P<0.00.

Table (3): Illustrates that there is a significant relation between total nurses' knowledge mean score, experience, and level of education before and after implementation of the educational program < 0.0.

Results

Table (1): Frequency and percentage distribution of Demographic characteristics of Nurses (n=70)

Items	No	%
Sex		
Male	32	45.7
Female	38	54.3
Age		
18-35	46	65.7
36-50	18	25.7
51-60	6	8.6
Level of Education		
Nursing Diploma	5	7.1
Technical Institute	11	15.7
bachelor's degree	42	60.0
Master degree	10	14.3
doctoral degree	2	2.9
Marital Status		
Single	38	54.3
Married	30	42.9
Widow	2	2.9
Years of Experience		
Less than 2 years	20	28.6
3-10 years	41	58.6
More than 10 years	9	12.9

Table (2): Comparison Between Mean Scores of Nurses' Knowledge Regarding VEXAS Syndrome Pre and Post-Implementation of the Educational Program (n=70)

Items	Pre-test		Post-test		Mean DF	T	P. Value
	Mean±SD	Mean%	Mean±SD	Mean%			
Definition	0.44±0.58	22.1	1.5±0.56	75.0	-1.06	-10.98	<0.001**
Causes	0.87±0.66	43.6	1.73±0.48	86.4	-0.86	-8.81	<0.001**
Symptoms	0.21±0.41	21.4	0.83±0.38	82.9	-0.61	-9.16	<0.001**
Lab test	1.17±0.98	29.3	2.96±0.58	73.9	-1.79	-13.17	<0.001**
Treatment	1.16±0.75	38.6	2.57±0.71	85.7	-1.41	-11.39	<0.001**
How to differentiate	0.33±0.61	16.4	1.16±0.65	57.9	-0.83	-7.78	<0.001**
Complication	0.73±0.64	36.4	1.69±0.53	84.3	-0.96	-9.71	<0.001**
Manifestation	1.93±1.74	32.1	4.83±0.88	80.5	-2.90	-12.44	<0.001**
Ng intervention	0.47±0.5	47.1	0.86±0.35	85.7	-0.39	-5.26	<0.001**
Total nurses' knowledge	7.31±4.04	31.8	18.11±1.75	78.8	-10.80	-20.53	<0.001**

- Paired T-Test Quantitative Data Between the Two Groups
*Significant Level at P Value < 0.05, **Significant Level at P Value < 0.01

Figure (1): Frequency Distribution of Knowledge Level about Nurses' Knowledge Regarding VEXAS Syndrome Before and After Educational Program (n=70)

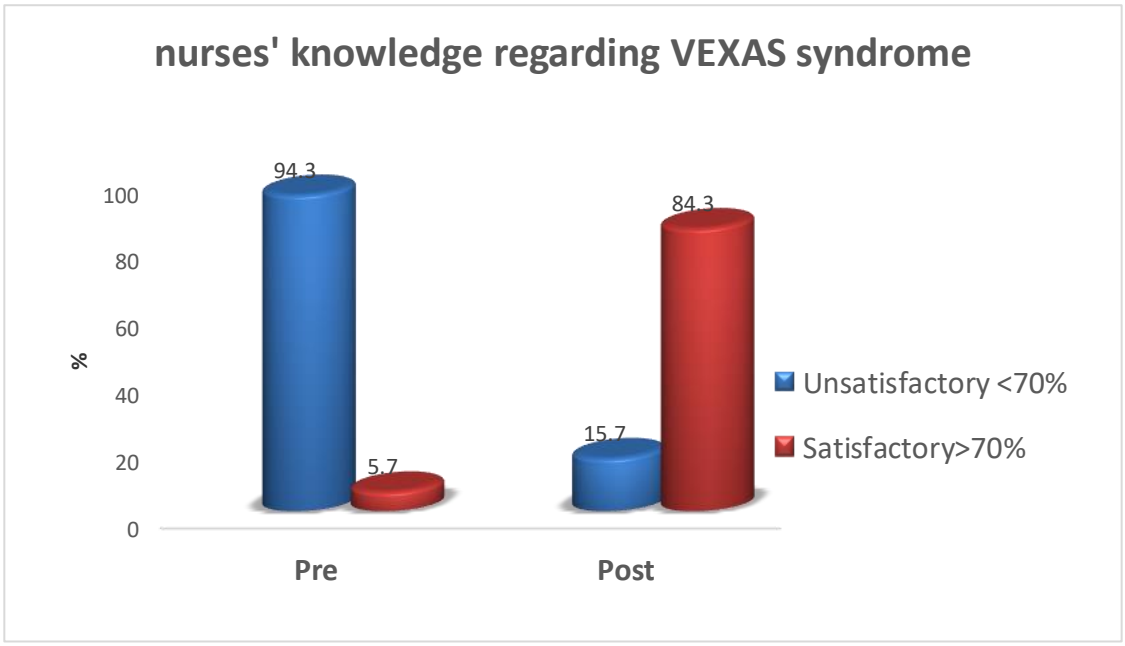

Table (3): Relationship Between Nurses' Knowledge Regarding VEXAS Syndrome and Their Demographic Data Before and After the Educational Program (n=70)

	N	nurses' knowledge regarding VEXAS syndrome			
		Pre-test		Post-test	
		Mean±SD	Range	Mean±SD	Range
Sex					
Male	32	6.88±3.25	2-17	18.16±1.55	14-21
Female	38	7.68±4.61	2-11	18.06±1.98	14-21
Test Used				P.V=0.822	
Age					
18-35	46	7.91±4.12	2-17	18.52±1.68	14-21
36-50	18	6±4.1	2-17	17.11±1.64	14-20
51-60	6	6.67±2.25	4-9	18±1.55	17-20
Test Used				P.V=0.013*	
Level of Education					
Nursing Diploma	5	10.8±1.64	8-12	17.2±0.84	16-18
Technical Institute	11	5.64±3.23	2-11	17.64±1.36	14-19
bachelor's degree	42	7.12±3.96	2-17	18±1.83	14-20
Master degree	10	7.9±5.43	2-17	19.4±1.71	16-21
doctoral degree	2	9±0	9-9	19±0	19-19
Test Used				P.V=0.04*	
Marital Status					
Single	38	6.92±4.28	2-17	18.24±1.67	14-20
Married	30	7.7±3.85	3-17	18.03±1.9	14-21
Widowed	2	9±0	9-9	17±0	17-17
Test Used				P.V=0.595	
Years of Experience					
Less than 2 years	20	6±4	2-12	18.1±1.74	14-20
3-10 years	41	8.29±4.07	2-17	18.24±1.62	15-21
More than 10 years	9	5.78±2.82	2-9	17.56±2.35	14-20
Test Used				P.V=0.01*	

- For quantitative data between the two groups, use an independent T-test. For quantitative data between three or more groups, use the one-way Anova T-test. *Level of significance at P < 0.05, **Level of significance at P <0.05

Discussion

Systemic inflammation and hematologic abnormalities are the hallmarks of vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic syndrome (VEXAS), a recently identified severe illness that primarily affects adult males. Critical care clinicians' knowledge of VEXAS syndrome is still extremely low, despite its significant influence on patient outcomes. This frequently results in delayed identification, diagnosis, and start of appropriate treatment. Managing VEXAS syndrome is extremely difficult due to the severe inflammation, organ dysfunction In fact, there have been reports of severe morbidity and mortality from this illness. Up to 40% of patients died, according to early accounts; later research has shown mortality rates as high as 50% **(Satoh et al., 2025).**

vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic syndrome (VEXAS), a novel hemato-inflammatory condition brought on by a somatic mutation in the UBA1 gene. Significant advancements have been achieved in comprehending the pathophysiological mechanisms, defining the epidemiology, describing the clinical phenotype, and discovering novel variations and promising treatments since Beck's initial description at the end of 2020. An international council of specialists now conducts recommendations for the diagnosis and treatment of VEXAS syndrome **(Khitri et al., 2024).** This study aims to evaluate the Effect of implementing educational program regarding vacuoles, E1-enzyme, X-linked, auto-inflammatory and somatic syndrome on nurses' knowledge in intensive

The study findings related to nurses personal and socio-demographic characteristics shows that more than half of the studied nurses were females and the rest were males. This agrees with **(Ou et al., 2021),** who stated that Respondents were predominantly female (99.4%). In researcher Opinion this is due to the historical background that was supporting that nursing is a female dominated profession and cultural association of nursing with femininity.

Concerning the age, about two thirds of the studied nurses was in the age group (18-35) years old, one quarter in the age group of (36-50) years old and the minority in the age group (51-60) years old. This agrees with Jooly,2025 who reported that the majority are aged between 20-35 years. Also agrees with **(Abdelmonem et al., 2018a)**, who reported that the studied nurses were in age group equal or much less than 25 years.

Regarding the studied nurses' qualifications, about two thirds of them had a bachelor's degree in nursing, and less than one quarter of them graduated from a nursing institute. This agrees with **(Ou et al., 2021),** who reported that most of the studied nurses were junior college graduates, and the rest were university graduates or above. The finding disagrees with **(Abdelmonem et al., 2018a**) , who reported that about half of the studied nurses were diploma nurses. The researcher sees that the presence of about two thirds of the studied nurses had bachelor's degrees in nursing due to the importance of critical situations faced by ICU nurses, which require highly qualified nurses who possess bachelor's degrees in nursing and higher degrees.

Regarding years of experience, nearly half of the studied nurses worked in ICU for more than three years; this agrees with (**Jaleta et al., 2021b)**, who reported that about half of the studied nurses had about five years of experience in ICU work. In the researcher's opinion, ICU nurses work with advanced technology and equipment requiring good clinical experience. Also, ICU patients require close monitoring and complex care, making the experience crucial for recognizing subtle changes.

According to the study's findings, nurses' post-test knowledge scores of VEXAS syndrome were considerably higher than their pre-implementation scores. This concurs with (**Salem FA, 2024**), who reported that there is a significant improvement in overall knowledge after implementation of the educational program. Also agree with **(Bakhit et al., 2024)**, who reported that the teaching program significantly improves nurses' knowledge and skills about patients with myasthenia gravis. These results are due to the use of good illustrating methods, as the video shows, and discussion.

The study shows that once the educational program was implemented, more than three quarters of the nurses had satisfactory knowledge, although the majority had unsatisfactory knowledge on the pretest. This is consistent with the findings of **(Abdelmonem et al., 2018b),** who reported that many nurses had inadequate expertise prior to the teaching program as opposed to the post-teaching program. Likewise, concur. This is consistent with (**Jolly, 2025),** who found that approximately 75% of the nurses in the study had inadequate knowledge of Guillain-Barré syndrome. In the researcher's opinion, the increase in the percentage of satisfactory level of knowledge of the ICU return is due to many causes. From the side of the nurses, this is due to their feeling of interest in knowing about the new issues; most of them have higher education, and many of them continue postgraduate education. From the other side, the explanation during teaching was supported with media, such as video shows.

The study's findings demonstrate a strong correlation between the mean score of total nurses' knowledge, their educational attainment, and their years of experience both before and after the training program was put into place. This is consistent with **(Jkhlab et al., 2025),** who stated that the study highlighted a strong positive association between demographic data such as years of experience and educational attainment. However, our study found that nurses' knowledge levels are positively correlated with their age. in keeping with **(Jaleta et al., 2021b),** who found a correlation between education level and good knowledge. In contrast, **Salem FA, 2024** found that sex and age had a substantial impact on knowledge score. From the researcher's point of view, educational level is an important factor in improving knowledge level regarding any new issue, as nurses with higher education have a good theoretical base that helps them to understand easier.

Conclusion

The majority of studied nurses had a satisfactory knowledge level, with a significant improvement in the total mean scores after implementation of the educational program.

Recommendations

Based on the findings of this study, the following recommendations are made:

Further multiple studies on a wide range of ICUs in many hospitals should be made for spreading the awareness among ICU nurses to help them in dealing with the newly arising cases of this disease.

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