

Effect of Health Educational Program on Improving Knowledge and Practices of Caregivers of Children with Xeroderma Pigmentosum

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Abstract: Background. Xeroderma pigmentosum (XP) is a rare autosomal recessive hereditary disease characterized by hyperphotosensitivity. **Aim.** This study aims to evaluate the effect of a health educational program on improving knowledge and practices of caregivers of children with xeroderma pigmentosum. **Subjects and Methods.** A quasi-experimental design with pre/post intervention assessments is utilized in this study with a purposive sample of 50 caregivers from 21 families of 30 children with a diagnosis of XP from the Pediatric Dermatology and Oncology outpatient clinics in the Children's Hospital, Ain Shams University in Cairo; the Dermatology outpatient clinic, Cairo University Hospital in Cairo; the Oncology Center, Mansoura University (OCMU) in Mansoura, and the Oncology Center, Tanta University in Tanta. **Results.** There were highly statistically significant differences between mean scores of pre/post test $p < 0.001$ in total mean scores of knowledge about XP that 38.13% of caregivers had poor scores, which improved to 71.98% good scores after implementation of the program. Regarding to practices, 42.45% of caregivers had poor practices, which improved to 76.78% that indicated improvement after providing of the educational program. **Conclusions and Recommendations.** The study demonstrated that implementation of an educational program had a significant positive impact on caregivers' knowledge and practice. The developed educational program should be implemented on a wider scale of nursing care and for longer time to assess long-term impact in further studies.

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Introduction

Xeroderma pigmentosum (XP) is a rare autosomal recessive hereditary disease characterized by hyperphotosensitivity, deoxyribonucleic acid (DNA) repair defects and a predisposition to skin cancers (Amr, Messaoud, El Darouti, Abdelhak, & El-Kamah, 2014). Xeroderma pigmentosum disorder is characterized by photosensitivity, pigmentary changes, premature skin aging and malignant tumour development due to cellular hypersensitivity to ultraviolet radiation resulting from a defect in DNA repair (Marwah, Garg, Chhabra, Dayal, & Sen, 2011).

Although XP is ultimately fatal, life can be prolonged by paying strict attention to simple preventive measures to minimize sun exposure. The patient should undergo regular checkups for early detection and treatment of any malignancies that may occur to reduce the mortality (Pradhan, Padhye, Malla, & Karki, 2003). The result is patients with XP are at a high risk for developing skin cancers, such as basal cell carcinoma; for this reason, the main presenting features of XP are photosensitivity, skin changes occur first over the areas most exposed to light, initially on the face (Webb, 2008).

Nursing educational programs for patients, caregivers and the public, about the signs and symptoms of XP is critical; as this could lead to earlier recognition

of the disease and subsequent earlier diagnosis. Therefore, the earlier protective measures can be undertaken to protect the patient from ultraviolet light, which can be helpful to avoid the catastrophic sequels of the disease (Digiovanna, & Kraemer, 2012).

Furthermore, Bradford *et al.* (2011) stated that, the diagnosis can often be made in the first years of life, about 60% of affected children demonstrated acute sun sensitivity (severe sunburn with blistering or persistent erythema on minimal sun exposure). Whereas, the remaining affected children did not burn easily but developed marked freckle-like pigmentation. These unusual freckles, when present on the face before age two years, are typical of XP and rarely seen in children with normal DNA repair mechanisms (Sethi *et al.*, 2013). Moreover, patients with XP have ocular abnormalities, which are caused by UV induced DNA alteration to epithelial cells of the conjunctiva, the cornea, and the eyelid (Feller *et al.*, 2010). Continued sunlight exposure may result in severe keratitis, leading to corneal opacification and vascularization, photophobia is often present and may be associated with prominent conjunctival injection, hyperpigmentation of the eyelids, loss of eyelashes, and in neoplasms (epithelioma, squamous cell carcinoma, and melanoma) (Hasan & Khan, 2011).

Xeroderma pigmentosum occurs with high frequency in Egypt and a continuation of our field studies has identified representatives of the 3 major complementation groups A, C, and variant. Group A patients, with one exception, showed very early onset of sun sensitivity and development of skin cancers, and microcephaly and mental retardation. Group C patients showed a slightly slower onset of sun sensitivity and had no central nervous system disorders. The variant type showed later onset of sun sensitivity and no skin cancers evident at the time of observation. No sun sensitivity was observed (Cleaver, Zelle, Hashem, El-Hefnawi, & German, 1981).

In a very recent study, Amr *et al.* (2014) reported that, all explored patients on the mutational analysis in Egyptian patients with XP-A, presented severe neurological abnormalities and have mutations located in the DNA binding domain. Meanwhile, there is a close relationship between the clinical features that ranged from severe to mild form and the mutational site in XPA gene.

However, Messaoud *et al.* (2010) discussed that, certain populations have a higher prevalence for example, North Africa including Tunisia, Algeria, Morocco, Libya, and Egypt, and the prevalence is increased, especially in communities in which consanguinity is common, genetic relatedness between individuals, XP has been found in all continents and across all racial groups. Consistent with autosomal recessive inheritance, males and females are similarly affected. Estimates suggested an incidence in North Africa and the Middle East, where there is a high level of consanguinity, is substantially higher (Lehmann, McGibbon & Stefanin, 2011). Although Mediterraneans seem to share common skin type and are subjected to similar environmental factors, they still represent a genetic and socioeconomic diversity and pigmentary disorders of XP in the Mediterranean area (El-Mofty, Esmat, & Abdel-Halim, 2007). So that, as part of a routine XP-related checkup, nursing staff and health care professionals should check skin of the child carefully, and should be willing to discuss any concerns the children and their caregivers might have about XP disease.

Magnitude of the Problem

Although there is no cure for XP, increased awareness and crucially early diagnosis, followed by rigorous protection from daylight and careful patient management, can dramatically improve the quality of life and life expectancy of affected individuals. A major challenge for the future is to increase understanding of the etiology of the neurological problems associated with the disorder in some individuals (Lehmann *et al.*, 2011). This should enable nurses on the development of health education program for caregivers, to assist them in developing ability to alleviate the symptom burden

during caring of their children suffering from xeroderma pigmentosum and protect them from any damage that occurs to the skin, eyes and nervous system from sun exposure is permanent and irreversible.

Aim of the study

This study aims to evaluate the effect of a health educational program on improving knowledge and practices of caregivers of children with xeroderma pigmentosum, through:

1. Assessment of caregivers' knowledge and practices about xeroderma pigmentosum.
2. Planning and implementing a health educational program according to the patients' needs.
3. Evaluating the health educational program outcomes.

Hypothesis

The study hypothesized that providing a health educational program will improve knowledge and practices of caregivers for children with xeroderma pigmentosum.

Subjects and Methods

Design

A quasi-experimental design with pre/post intervention assessments is utilized in this study.

Setting

The study was conducted at the Pediatric Dermatology and Oncology outpatient clinics in the Children's Hospital, Ain Shams University in Cairo; the Dermatology outpatient clinic, Cairo University Hospital in Cairo; the Oncology Center, Mansoura University (OCMU) in Mansoura, and the Oncology Center, Tanta University in Tanta, which all admit children with xeroderma pigmentosum conditions.

Subjects

A purposive sample of 50 caregivers, from 21 families of 30 children with a diagnosis of XP was recruited from the mentioned settings. All caregivers were family members of the involved children with XP and they were willing to participate in the study. Exclusion criteria included caregivers of children under treatment from severe skin or respiratory infections or any other infectious diseases. All caregivers met inclusion criteria.

Although this study meant caregivers and their children to join in the research sample, but most of the children were in health conditions tough physically and neurologically, so do not permit them to receive health education program or absorb the information and apply the desired practices. Only 9 of the children patients who aged more than 15 years had participated in the study sample as self caregivers because they were the ones who take care of themselves.

Data Collection

Two different tools were designed and used. They included knowledge questionnaire sheet (pre/post formats), and observation checklist (pre/post formats).

Questionnaire Sheet (Pre/post Format).

Socio-demographic data characteristics of the patient and caregiver such as age, gender, duration of illness, level of education, and attended training courses previously about XP, knowledge questionnaire (pre/post format). It is used for assessment of caregivers' knowledge about principal landmarks of xeroderma pigmentosum including, identification of xeroderma pigmentosum; progress stages; signs and symptoms; management of XP; complications; safety precautions outdoor and indoor activities; and inheritance and genetic counseling.

Observation Checklist (Pre/post Format).

This is used to assess each caregiver's practices during daily aspects of care provided for children suffering from XP. It consisted of five directions with a total of 11 checklists as follows:

Direction 1: General assessment:

- (a) Inspection the lesion from head to toes;
- (b) Measuring vital signs of temperature, respiration patterns, and pulse;
- (c) Reviewing growth aspects such as head circumference, height, and abdominal girth.

Direction 2: General inspection for:

Eye disorders, neurologic abnormalities and skin illness:

- (a) Eye inspection of:

Bright flashes of light, changes in vision, blind spot bordered by shimmering light, spots in visual field (floaters), double vision (diplopia), check for clouding of the cornea, tumors near the eye, pain or other problems;

- (b) Neurologic abnormalities inspection as following:

Fainting or passing out; shaking that you can't stop or seizures; dizziness; loss of balance or lack of coordination; alterations in consciousness; memory disorder; headache or any other disorders;

- (c) Skin illness including:

The location (such as arms, head, legs), symptoms (pruritus, pain), duration (acute or chronic), arrangement (solitary, generalized, annular, linear), morphology (macules, papules or vesicles), and color (red, blue, brown, black, white or yellow).

Direction 3: Applying eye and skin care:

- (a) Eye care;
- (b) Skin care.

Direction 4: Medication administration (Oral, eye and skin) medications:

Use the five rights of medication administration:

- (a) Preparation;
- (b) Administration of the prescribed medications.

Direction 5: Assessment of stress problems:

Perceived Stress Scale (PSS) (Cohen, Kamarck, & Mermelstein, 1983): It is classified into 3 categories as follows:

- (a) Low stress,

- (b) Moderate stress, or
- (c) High perceived stress

Educational Program

The researcher developed and implemented an educational program about xeroderma pigmentosum for caregivers. The program phases were:

Exploratory phase: Baseline assessment for caregivers had been done before the development of the program content.

Validity and reliability: During the program construction phase, the content of pediatric xeroderma pigmentosum educational program was provided for caregivers of children patients, which based on the assessed caregivers' needs. In addition, after reviewing related literature (e.g., Lashley, 2005; Tomlinson & Kline, 2005; Lei, 2007), a group of experts reviewed the program and evaluated the program contents included the assessment tools; they were Oncology and Dermatology Consultants, and the Head Nurses of the Pediatric Oncology and Dermatology Units that refer to the extent to which assessments are consistent.

Implementation phase: A series of four sessions were offered along 2 days/ week (Saturdays and Thursdays) in the morning shift, over a 16-week period, according to the caregivers' availability, with a total of 4 hours for each child and his/her family caregivers. One hour was allocated to theoretical content, with two time-breaks for five minutes each. This part covered the required knowledge about using the principal landmarks about XP.

The practical part of the program was provided through two sessions, one hour each, for a total of 2 hours. This part covered training on general assessment of vital signs, monitoring the lesion: from head to toes and reviewing growth aspects such as head circumference, height, and abdominal girth. In addition, general inspection of eye disorders, neurologic abnormalities and skin illness, which were done during applying of the eye and skin care, to save the time for further demonstrations and answering vague questions of caregivers. More directions that are practical were related to administration of oral, eye and skin medications.

Furthermore, assessment of stress problems by using the Perceived Stress Scale (PSS), which is classified into 3 categories, they were low stress, moderate stress, or high perceived stress (Cohen *et al.*, 1983), pre/post-tests were applied before and after the XP program intervention.

During the implementation stage of this educational program, participant caregivers were clarified about the objectives of the study, and a simple introduction. Then the program was initiated by the theoretical part, and followed by the practical part (The study period spanned from June to December 2013).

Teaching strategies included small group

discussions; in fact, most of the groups were caregivers from the same family, and they were all caring of their child with XP, who needs to strenuous efforts of all members of the family to care for them. In addition, demonstrations, and remonstrations were done and caregivers had the chance for performing each practice and for correcting of mistakes on the spot.

Appropriate visual media used included printed and illustrated colorful, handouts and posters. In addition, real equipment needed for assessing and managing XP such as; a thermometer, a watch, designed colorful charts, pencils for measuring vital signs, assessing the child physically, and reviewing growth aspects. In addition, gauze, materials, and solutions required for wound care, were actually used for the training to ensure maximum efficacy.

For practicing the administration of medications, on one hand, the researchers used either the prescribed eye drops or ointments, or acting role-play during ophthalmic medications, and washing and drying hands. On the other hand, the researchers brought real samples of creams, foams, gels, lotions and ointments for practicing caregivers on different types of topical applications.

The researchers introduced packets of tablets, capsules, powders or granules and water, liquid medications or syrups in addition to other equipment required for educating participants about correct preparation and administration and monitoring patients with XP after taking medications.

Management of stress started with identifying the stress levels, causes, and symptoms of stress. In addition, sessions were tailored towards meeting caregivers' individual needs to overcome their stress from their children's XP disease and its sequences.

Thus, SSP questions were used for appraisal of feelings and thoughts of caregivers during the last month, this scale classified stress into 3 categories of low stress, moderate stress, or high perceived stress. Then, the researchers spent one-hour session for demonstrating caregivers about the process of coping strategies included, light exercises for managing anger when present, talking to others about physical and psychological problems, and taking time for regular relaxation practices, and sharing their experiences of stress and problems together.

Evaluation phase: The main target of this study focused on evaluating the caregivers' knowledge, practices and stress towards XP; that were assessed after the program using the same tools of the pre-program intervention.

The questionnaire sheets were completed by caregivers of children with XP in the setting area of the mentioned units. According to the designed model key answers, the knowledge of caregivers was categorized into two categories; either "unsatisfactory" when the

total score obtained was less than 60%, or "satisfactory" when 60% or more from the total score of the provided questionnaire.

The observation checklists were completed for assessment of caregivers' practices during preparing, and applying practices related to assessing and managing their children with XP health problems. The practices of the caregivers were categorized into "adequate" once they had 70% or more of the total scores or "inadequate" when less than 70%.

Perceived Stress Scale (PSS) is used to reflect the reaction of stress. Caregivers were asked to answer the 10 questions in this scale about feelings and thoughts during the last month. Thus, rating stress is obtained by reversing the scores on the four positive items, 0=never; 1=almost never; 2=sometimes; 3=fairly often; and 4=very often. Accordingly, a total score of PSS is obtained by summing together the scores for the ten questions, which is calculated and interpreted as follows: scores ranging from 0-13 would be considered "low stress", scores ranging from 14-26 would be considered "moderate stress", and scores ranging from 27-40 would be considered "high perceived stress" (Cohen *et al.*, 1983).

Knowledge, practices and stress assessments were evaluated twice for each participant caregivers before and after an exposure to the educational program about XP.

Pilot Study

Five piloted caregivers were asked to answer the questionnaire. They were observed during applying practices which managing their children with XP to appraise the applicability of the tools regarding their knowledge and practices. Modifications such as adding children who were caring for themselves and aged more than 12 years old; they were added as self caregivers according to the pilot results. In addition, stress assessment and management was added to the program intervention.

Ethical Considerations

Ethical issues in this study involved the assurance of confidentiality and anonymity for the participants. All caregivers had received a verbal explanation about the purpose and process of the study; and they were allowed to withdraw from the study whenever they wished without giving any reasons. Requests for permission to conduct the study were obtained from Manager of the hospital, and the Chairperson of the Pediatric Dermatology and Oncology outpatient clinics involved in the study. The same procedures were applied to caregivers and their children after obtaining informed consent/assent from them.

Statistical Analysis

Data were fed to the computer and analyzed using Statistical Package for the Social Science (SPSS) version 20.0 Qualitative data were described using

number and percent. Quantitative data were described using mean and standard deviation for normally distributed data. Comparison Values from caregivers' related knowledge, and practices were analyzed, then compared and categorical variables were tested using the Wilcoxon signed ranks test. Significance test results are quoted as two-tailed probabilities. Significance of the obtained results was judged at the 5% level.

Results

Table (1): Shows that the mean age of children with xeroderma pigmentosum was 11.03 ± 4.31 and 53.3% of them were in the age group of ≤ 10 years. Regarding sex, 60.0% of the study sample were males, 40.0% of them had read and write.

Table (2) shows that the mean age of caregivers was 35.5 ± 11.56 and 38.0% of them were in the age group of 35 - 45 years. Regarding kinship with the patient child, 58.0% of the study sample was mothers; while 74.0% of them did not any receive any training courses previously.

Table (3) reveals that percentages of caregivers' knowledge about xeroderma pigmentosum disease at pre/post program. It is obvious that most of the studied sample pre program had poor knowledge about identification of XP (86.0%), signs and symptoms (82.0%), progress stages (90.0%), environmental control of the sun exposure (80.0%), complications (86.0%), management of XP (82.0%), safety precautions of outdoor activities (94.0%), Safety precautions of indoor activities (86.0%), and inheritance and genetic counseling (88.0%) respectively. However, good knowledge of caregivers improved after implementation of the program (38.0%, 38.0%, 36.0%, 44.0%, 26.0%, 36.0%, 28.0%, 40.0%, 32% & 44% respectively). There were highly statistically significant differences between pre and post knowledge related to poor, average and good knowledge ($P < 0.00$).

Table (4) indicates that majority of mean scores of caregivers' practices were poor before implementation of the educational program especially in relation to neurologic abnormalities (88.0%). Conversely, after implementation of the program 44.0% of them had reached maximum good scores. Moreover, the pre test showed that most of caregivers 86.0% had poor scores regarding specific examinations of eye disorders, but the post test showed some improvement just 30% in this important skill. Caregivers did not have too much improvement from 78.0% pre program in measuring vital signs, skin illness, eye care, and preparation of (oral, eye and skin) medications to be 32%, 38%, 36% and 40% respectively. There were highly statistically significant differences in mean scores of all practices of caregivers

in caring for their children with XP ($P < 0.00$).

Regarding caregivers' scores in managing psychological stress problems pre/ post program implementation, **Table (5)** reveals that only one tenth of the caregivers had mild stress at the preprogram, their scores improved at the post-test and 62% of them succeeded to manage stress. The pre-test shows that 32% of the caregivers had high scores according to the rating categories of Perceived Stress Scale (PSS), compared to 18% at the post-test. Overall statistical significance difference after the educational program intervention (negative mean ranks=12.5& positive mean ranks=19.37) indicating an improving of caregivers' stress scores toward their children's illness compared to their stress scores at the pre-test ($P < 0.00$).

Table (1): Percentage distribution of the children under study according to their socio-demographic characteristics (n=30).

Socio-demographic characteristics	n	%
Age (in years)		
<10	16	53.3
10-15.	5	16.7
>15	9	30.0
Mean \pmSD	11.03 \pm 4.31	
Gender		
Male	18	60.0
Female	12	40.0
Level of Education		
Illiterate	9	30.0
Read and write	12	40.0
Preparatory school	8	26.7
Secondary school	1	3.0

Table (2): Percentage distribution of the caregivers according to their socio demographic Characteristics (n=50).

Characteristics of caregivers	n	%
Age (in years)		
<25	9	18.00
25 - < 35	15	30.00
35 - < 45	19	38.00
45 +	7	14.00
Mean \pmSD	35.5 \pm 11.56	
Kinship with the patient child		
Mother	29	58.00
Father	12	24.00
Grandfather/grandmother	6	12.00
Self	3	6.00
Attended training courses previously		
No	37	74.00
Yes	13	26.00

Table (3): Mean scores of caregivers' knowledge regarding XP disease pre/ post program (n=50).

Caregivers' Knowledge		Pre			Post			Mean Rank(post-pre)		Wilcoxon Signed Rank Test	
		Poor	Average	Good	Poor	Average	Good	Negative Ranks	Positive Ranks	Z	*P- value
Identification of XP	n	43	7	0	9	22	19	39.50	52.47	-3.30	0.001
	%	86	14	0	18	44	38				
Signs and symptoms	n	41	9	0	7	24	19	16.00	26.20	-4.91	0.00
	%	82	18	0	14	48	38				
Progress stages	n	45	5	0	15	17	18	7.50	16.79	-4.95	0.00
	%	90	10	0	30	34	36				
Environmental control of sun exposure	n	40	10	0	9	19	22	0.00	18.50	-5.40	0.00
	%	80	20	0	18	38	44				
Complications	n	43	7	0	9	28	13	0.00	18.00	-5.37	0.00
	%	86	14	0	18	56	26				
Management of XP	n	41	9	0	14	18	18	10.00	18.75	-4.82	0.00
	%	82	18	0	28	36	36				
Safety precautions of outdoor activities	n	47	3	0	13	23	14	0.00	18.50	-5.45	0.00
	%	94	6	0	26	46	28				
Safety precautions of indoor activities	n	43	7	0	6	24	20	0.00	20.50	-5.70	0.00
	%	86	14	0	12	48	40				
Inheritance and Genetic counseling	n	41	9	0	13	21	16	11.50	17.68	-5.08	0.00
	%	82	18	0	26	42	32				
Inheritance and Genetic counseling	n	44	6	0	12	16	22	0.00	17.50	-5.26	0.00
	%	88	12	0	24	32	44				

*P- value Statistically Significant at (P <0.000).

Table (4): Distribution of mean scores of caregivers' practices caring for their children with XP pre/ post program implementation (n=50).

Practices of Caregivers		Pre			Post			Mean Rank(post-pre)		Wilcoxon Signed Ranks Test	
		Poor	Average	Good	Poor	Average	Good	Negative Ranks	PositiveRanks	Z	*P-value
Direction 1: General assessment											
Inspection the lesion from head to toes	n	43	7	0	13	16	21	13.50	19.49	-4.92	0.000
	%	84	18	0	26	32	42				
Measuring vital signs	n	39	11	0	13	21	16	10.50	20.25	-5.46	0.000
	%	78	22	0	26	42	32				
Reviewing developmental aspects	n	40	10	0	14	20	16	10.50	18.18	-4.70	0.000
	%	80	20	0	28	40	32				
Direction 2: General inspection											
Eye disorders	n	43	7	0	16	19	15	11.00	20.23	-5.05	0.000
	%	86	14	0	32	38	30				
Neurologic abnormalities	n	44	6	0	10	18	22	10.00	15.50	-4.95	0.000
	%	88	12	0	20	36	44				
Skin illness	n	36	13	0	9	22	19	8.00	17.79	-5.11	0.000
	%	78	22	0	18	44	38				
Direction 3: Protecting eye and skin measures											
Eye care	n	39	11	0	12	20	18	11.50	18.91	-5.05	0.000
	%	78	22	0	24	40	36				
Skin care	n	40	10	0	9	22	19	11.50	19.94	-5.21	0.000
	%	80	20	0	18	44	38				
Direction 4: Medication administration (Oral, eye, skin) medications											
Preparation	n	39	11	0	8	22	20	13.00	20.18	-5.47	0.000
	%	78	22	0	16	44	40				
Administration	n	41	9	0	10	23	17	12.50	19.18	-5.32	0.000
	%	82	18	0	20	46	34				

*P- value Statistically Significant at (P <0.000).

Table (5): Distribution of caregivers' scores in managing psychological stress problems pre/ post program implementation (n=50).

Rating categories		Pre			Post			Mean Rank(post-pre)		Wilcoxon Signed Ranks Test	
		Mild	Moderate	High	Mild	Moderate	High	Negative Ranks	Positive Ranks	Z	*P-value
Perceived Stress Scale (PSS)	n	11	23	16	31	19	9	12.50	19.37	-5.12	0.000
	%	22	46	32	62	38	18				

*P- value Statistically Significant at (P <0.000).

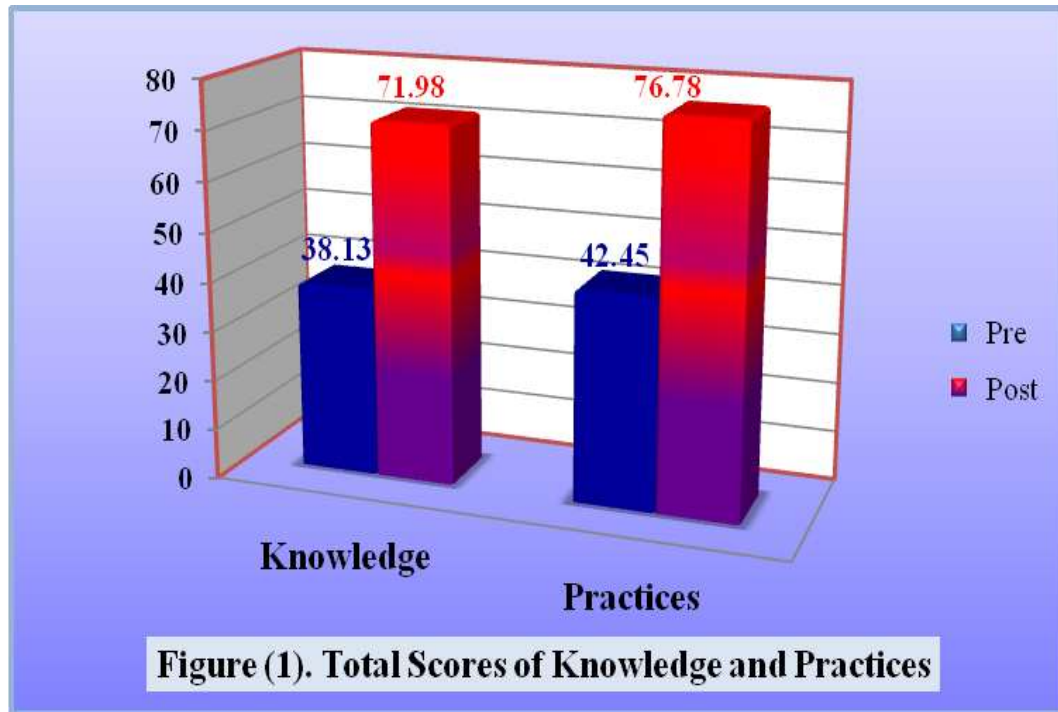


Figure (1): Illustrates changes in total mean scores of knowledge that 38.13% of caregivers had poor knowledge about XP, improved to 71.98% good knowledge after implementation of the program. Regarding to practice 42.45% of caregivers had poor practice, which improved to 76.78% after implementation the program.

Discussion

Children of the ‘night’ and ‘moon children’ are examples of the names used to describe those with xeroderma pigmentosum (XP), a severe, lifelong, genetic DNA repair disorder. These children with XP live their life in potential fear of daylight, which has the ability to harm them (Turner, Mullard, Fassihi, & Sarkany, 2013). From this point of view, nurses can play a crucial role in improving health and supporting those children and their caregivers. Therefore, for the first time ever in Egypt, this study is done by providing an appropriate health educational program for caregivers of children with XP for improving their knowledge, practices and to meet their requirements through nursing support.

The current study findings showed that, before the educational program, none of the studied group of caregivers had a good background about the identification of XP or the signs and symptoms of the disease. This could be attributed to the absence of educational materials about XP in Arabic language in

any of the involved hospitals. In addition, it could be due to lack of knowledge, lack of experience of nurses because XP is a rare disease, also, lack of time for dermatologists and nurses to provide patients and caregivers with proper information although they are the main source of information for those patients. These rationales are suggested not only for the poor scores about meaning of XP, but also to caregivers’ knowledge regarding all items related to XP disease at before implementation of the program. After implementation of the educational program, the caregivers became oriented that signs and symptoms of XP can initially be observed in the sun exposed areas of the skin and eyes in addition to atrophy, telangiectasia, skin tumors, and oral carcinomas could be other common manifestations (Zeynep, Efsun, & Firdevs, 2012).

The study findings indicated low knowledge scores before implementation of the educational program. This is particularly indicated in the cutaneous, ocular, and neurologic abnormalities’ complications and management of XP although these two areas were of

major critical for the decision of proper management. This finding might be related to the low educational attainment of caregivers as the majority of them were illiterates.

The most obvious and often important part of the educational program is about safety precautions of outdoor and indoor activities for avoiding exposure to sunlight. This study highlighted statistically significant differences in scores of caregivers. Pre-program, most patients and their caregivers were having difficulty to get good scores; whereas, post-program, most of them expressed good understanding. Firstly, safety outdoor precautions; by wearing protective clothing as long-sleeved shirts and full-length pants, shirts with collars, tightly woven fabrics that do not let light through. In addition, the use of wide-brimmed hats; and eyewear specifically made to screen out ultraviolet rays; applying sunscreens; avoiding going outdoors during daylight hours, and limiting outdoor activities to nighttime only are other precautions to be considered (Andersen *et al.*, 2010). In this regard, Lehmann *et al.* (2011) suggested that, when children are outside during daylight hours, exposed skin should be covered with sunscreen and lip balm. In addition, long trousers, long sleeves, and gloves should be worn together with a UV-resistant facemask. If the patient finds this unacceptable, a broad-brimmed hat or "hoodie" and wrap-around sunglasses should be worn.

Another concern for children XP and their caregivers was about restricting indoor activities, although only seven of caregivers had average scores before implantation of the educational program, they had got high scores indicating understanding about indoor restricted activities by avoiding the use of halogen or fluorescent bulbs indoors; these give off enough ultraviolet radiation to affect people with XP. In addition, protective film on windows that blocks ultraviolet rays to house and car windows, special eyeglass filters to block UV light, and avoiding the environmental triggers such as smoking, passive smoking and other allergens. In this respect, Digiovanna and Kraemer (2012) revealed standard precautions that, XP patients should limit their activities to the indoors or at night. If they are done outside, the patient must use appropriate sunblocks and cover with a broad-brimmed hat and long-sleeved clothing. Patients can also wearing National Aeronautics and Space Administration (NASA) -like suits when participating in outdoor activities. These suits serve as a head-to-toe physical skin barrier, fully protecting patients from UV radiation. In congruence with this, Turner *et al.* (2013) emphasized that, if protective measures are put in place early in life, some of the complications seen from XP, in particular skin cancers, can be reduced or avoided all together by reducing exposure of people with XP to ultraviolet (UV) radiation in normal everyday tasks.

After the program, considerable improvement was detected in patients' knowledge, which could be attributed to the face to-face theory and practice sessions given by the researchers together with leaflets and posters about XP.

On the other hand, caregivers have high knowledge scores regarding the progress stages of XP, even before implementation of the educational program. Moreover, caregivers realized that the first features are either extreme sensitivity to sunlight, triggering severe sunburn, or, in patients who do not show this sun-sensitivity, abnormal freckle-like pigmentation due to increased numbers of melanocytes, on sun-exposed areas. This is followed by areas of increased or decreased pigmentation, skin aging, and multiple skin cancers, if the individuals are not protected from sunlight. A minority of patients show progressive neurological abnormalities of varying severity (Lehmann *et al.*, 2011).

Concerning knowledge about inheritance and genetic counseling, in the present study, results revealed that caregivers were not oriented about the meaning of genetic disorders. However, post-program, they expressed a wide difference understanding about the importance of attending a genetic counseling and get psychological support. In congruence with this, Lehmann *et al.* (2011) emphasized that genetic counseling is essential for the affected families, to discuss about etiology, probability of occurrence in future pregnancies, increased likelihood of occurrence in communities in which consanguineous marriages are common, and feelings of isolation and concern about career prospects. Explicitly, the inheritance is autosomal recessive, where XP is suspected, siblings should be protected from UV light until XP can be excluded. If parents are considering further pregnancies, prenatal diagnosis is often possible (Webb, 2008).

The present study also assessed caregivers' practices related to caring of their children with xeroderma pigmentosum. The results indicated that after application of the educational program, statistically significant improvements were noticed in all areas of the first direction of related skills (general inspection). Furthermore, caregivers reached maximal scores at the post-test in the terms of monitoring the lesion from head to toes, and reviewing growth aspects. Certainly, the high scores of these skills were clearly; due to the fact that, one of the most important aspects of daily care for children is observing for any changes related to the disease on the child and following the child's growth and development. Microcephaly, delayed motor development, sensory neural deafness are commonly seen (Pathy *et al.*, 2005) in children with XP.

Although measuring vital signs are necessary for early detection of any abnormalities and asking for medical advices, a relative increases in the scores were

noticed in taking and understanding normal ratios of pulse, temperature and respiratory measurements, after application of the educational program. Caregivers were trained to take vital signs including assessment of temperature, pulse and respiration, and comparing them with the first time they were taken with the researchers. They became oriented that, vital signs should be taken for one full minute, and they are affected by a variety of internal and external factors, including many disease conditions, stress, pain, fatigue, and even exercise or any changes in the ill child daily rhythms.

The researchers gave much effort to demonstrate caregivers about taking pulse for the XP children, in any way caregivers who were illiterates couldn't count more than number ten, so that they couldn't progress in counting the pulse at all. Certainly, vital signs and other growth parameters were evaluated to both checking for abnormal values and providing physicians with new measurements. In a study of, Kyriacos, Jelsma, and Jordan (2011) suggested a new idea to record vital signs on a number of charts such as respiratory rate, temperature, heart rate and blood pressure, oxygen saturation and level of consciousness and urine output. Recoding was achieved by converting each recorded value (for example HR 50) for each observation time-point into a score and reporting it as normal (0) or having a low or high score of 1–3 on the review form, using the study criteria.

An illustrated coloured chart preliminary designed by the researchers, was easy for all caregivers even for the read and write and the illiterate ones, who had learned how to record the growth and developmental measurement. Where, there is a horizontal line located in the middle of the chart indicating the current measurement by the caregivers after the first demonstration and supervision of the researchers. Then, caregivers plotted on the chart, either above this line if the measurements were higher the present measures, or under the line if it was less the present one. Moreover, caregivers had learned to use the tape, which is marked by the researchers according to the initial degree of measurements. Here, they can draw using a pencil other future remarks on the measurement tape and record on the designed developmental chart.

After application of the educational program, statistically significant improvements were noticed in the caregivers' practices regarding specific assessment especially for eye disorders, which is very important for early detection and changes to develop proper intervention. Thus, Alfawaz and Al-Hussain (2011) emphasized that patients with XP who presented to a tertiary eye care center in Saudi Arabia had a high percentage of consanguinity in parents and late onset of ocular complications. In addition, XP behaved aggressively in both malignancy profile and visual outcome. Hence, children with XP must avoid exposure

to sources of UV light; key points of this study include avoidance of sun exposure at peak hours, patients must wear protective sun glasses and lenses, preferred UV absorbing eye glasses, and careful daily eye care and use of sunscreen. The XP demands protection of the patient from artificial light exposure (Oliveira, Elias, Barros, & Conceicao, 2003; Feller *et al.*, 2010). Moreover, eyes that are painful due to disorders of the cornea and eyelids, associated with UV- exposed portion of the eyes are conjunctiva, cornea, and lids, showing ophthalmologic abnormalities include photophobia, which is often present and may be associated with prominent conjunctival injection and loss of lashes. Atrophy of the skin of the lids results in ectropion, entropion, or in severe cases, complete loss of the lids (Brooks *et al.*, 2013). Thus, this program involved assessment of eyes abnormalities for careful protection of eyes as the mainstay of prevention in all XP children patients.

Successfully, caregivers became familiar with the practices related to routine measurement of head circumference and assessment, in addition to assessment of gait and speech and hearing testing can usually serve as a screen for the presence of XP-associated neurologic abnormalities. Unfortunately, Lehmann *et al.* (2011) stated that, the cause of the neurological problems is not fully understood, and there is no known way of preventing them. Nursing management of XP patients with neurological involvement can include early detection and visiting the child's physician for referral of hearing, physical and speech therapy and yearly testing of the patient's nervous system. Moreover, Bradford *et al.* (2011) reported that 25% of affected individuals had characteristic progressive neurologic manifestations that worsen slowly and may manifest later than the skin changes. Consequently, Lehmann *et al.* (2011) identified that there is a huge variation in clinical features, and the neurological abnormalities are dependent on exposure to sunlight, the complementation group, the precise nature of the mutation as well as unknown factors.

Another important finding is the statistically significant difference, after implementation of the educational program, in relation to practices of caregivers in applying specific assessment of any skin illness in children with XP. However, Tamura, DiGiovanna, and Kraemer (2010) advised that, XP affected children or their parents should be educated to look for abnormal pigmented lesions or the appearance of basal cell or squamous cell carcinoma. Furthermore, a family member who has been instructed in recognition of cutaneous neoplasms should examine children frequently. Of particular interest is that all participants seriously applied protective outdoor and indoor measures because they became oriented that sunny climates, outdoor living, smoking, poor protection from

sunlight will exacerbate the cutaneous features, resulting in multiple pigmentation changes, multiple skin cancers and early death. Conversely, less sunny climates, indoor living, pigmented skin, early diagnosis and good solar protection, can result in relatively mild skin features (Lehmann *et al.*, 2011).

Several attempts were done by the researchers to instruct caregivers about eye care for their children. Pre-program, none of the patients applied this practice correctly; most of caregivers didn't perform eye exercises and very few were using eye compresses. After completion of the program, the caregivers became more skillful in that compresses are to be applied over the eyes for 2-3 minutes with cold water for each eye alternatively. They also applied eye exercises as a vital step for increasing eye relaxation and decreasing stress, which resulted in children being gifted with a wider eye view and range of vision. In this respect, Lin, Shih, Hsiao, and Chen (2004) found that children do not complain of defective vision, and may not even be aware of their problem. They adjust to the poor eyesight by sitting near the blackboard, holding the books closer to their eyes, squeezing the eyes and even avoiding work requiring visual concentration. This warrants early detection and treatment to prevent permanent disability, and they concluded that the cause of the relative increasing severity of myopia among the school children was due to the onset of myopia at a very young age. Thus, to reduce the prevalence and severity of myopia, more attention should be paid to the eye care of pre-school children.

The present study analysis of patients' skin care practices showed that the majority of them did not practice topical irrigation therapy for cleaning open wounds correctly pre-program. Caregivers of XP patients utilized a syringe for open wounds in order to loosen debris, and they used gauze or cotton swabs well enough with caution as they can cause mechanical damage to new tissue and delays healing, and they were able to remove excess exudates. Explicitly, health care providers may neglect to help patients and their caregivers to practice techniques of skin care probably. In this regard, Oliveria *et al.* (2001) concluded that formal large-scale programs need to be developed in order to train nonphysician health care providers to successfully screen and detect early skin cancer. Moreover, post-program scores improved in regard to personal hygiene and complete bed bathing.

Another concern was related to the administration of medications based on the established educational program. This study finding highlighted that statistically significant difference was detected in scores of caregivers in practicing the five rights during medication administration; relative high scores were related to the application of topical medications. Distinctly, Zaghi and Maibach (2007) stressed that, topical medications differ

from many other types of drugs because mishandling them can lead to certain complications in an XP patient or administrator of the drug. Clearly, caregivers had high scores in the term of preparation and administration of ophthalmic medications after program intervention of the practical sessions. In addition, they practiced the oral medications adequately even before implementation of the program, which includes buccal, sublabial and sublingual administration of medications that come in various forms.

Despite management of stress is not a study target, but ethically, the researcher couldn't abandon this problem. The researchers started with identifying the true sources of stress, which were related mainly to the nature of XP as a rare illness. Moreover, the present study result revealed a high prevalence of stress problems among caregivers; pre-program, more than half of them had "moderate or high perceived stress" where, children and families were worried about deadlines of XP progress that is not as easy as it sounds. Obviously, the patients showed unhealthy ways of coping with stress such as over eating or under eating and taking out stress on others, being angry and outburst; it was easy to overlook those stress reactions. So that, the researchers shed light on stress-inducing thoughts, feelings, and behaviors.

At the beginning of stress management, the researchers urged caregivers to increase their awareness about stress levels, causes of stress and paying attention to their children's symptoms of stress. At the same time, the program intervention assisted caregivers in taking good care of their children's bodies as one of the coping strategies included, light exercises for managing anger when present, talking to others about physical and psychological problems, and taking time for regular relaxation practices were recommended by Artherholt and Fann (2012). After intervention with the coping strategies suggested by the present study program, findings revealed that SSP measures indicated more "low stress" among caregivers leading to more self-acceptance of their children illness, and a minority of them had "high perceived stress".

Successfully, all caregivers had proper understanding and adequate practice at the post-test in all knowledge and practice components, compared to their scores in the pre-test. In this respect, Lehmann *et al.* (2011) underscored the extreme importance of appropriately educating patients as well as actively protecting the skin from ultraviolet radiation for XP patients who must practice sun avoidance and whenever necessary use good sunscreen protection.

Conclusion

The study demonstrated that implementation of an educational program had a significant positive impact on caregivers' knowledge and practices and led to higher

educational and practical scores was related to the preparing and administration of topical and ophthalmic medications after program intervention of the practical sessions.

Recommendations

Based on the study findings, it is recommended that innovative methods of educational programs are needed to improve the knowledge and practices of XP patients and their caregivers.

Further studies are needed to address the reduction of the psychosocial problems associated with quality of life of patients affected with XP.

Xeroderma pigmentosum and with all other genetic disorders, it is recommended to provide genetic counseling and psychological support for the affected families, to discuss etiology, probability of occurrence in future pregnancies, increased likelihood of occurrence in communities in which consanguineous marriages are common.

Further research is required to identify the impact that age and developmental status have on the XP sufferers.

The developed educational program should be implemented on a wider scale of nursing care and for longer time to assess long-term impact in further studies.

Implications for nursing

Since the health educational program proved to be useful in improving knowledge and practices of caregivers of children with xeroderma pigmentosum, it could be implemented on the national level to improve the quality of care for these patients and their families.

Limitations of the study

We acknowledge that a longer period would be desirable but difficulty of transportation to reach those patients' homes, in addition to their expectation of giving them some tools or funding for this disease did not permit follow-up beyond this current study and results.

Conflict of interest

All the listed authors have agreed to the submission of the manuscript in its current form. The authors have no commercial affiliations, consultancies, stock or equity interests and patent licensing arrangements that could be considered to pose a conflict of interest regarding the submitted article.

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