

Effect of Educational Program on Mothers' Practices towards Caring for their Children Having Phenylketonuria

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Abstract

Background: Phenylketonuria (PKU) is an autosomal recessive inborn error of phenylalanine metabolism resulting from deficiency of phenylalanine hydroxylase (PAH). **Aim:** The objective of this study was to evaluate effect of educational program on mothers' knowledge, practices and behavior towards caring for their children having Phenylketonuria. **Methods:** A pre/post quasi experimental study was conducted on 128 mothers. **Settings:** The study conducted at the Metabolic Clinic, Department of Medical Genetics, Faculty of Medicine, Ain Shams University. **Tool:** Pre designed interviewing questionnaire consists of 4 parts: Part 1: Personal characteristics of mothers. Part 2: Mothers' knowledge about PKU. Part 3: Mothers' Reported Practice about PKU. Part 4: Mothers Reported Behavior. **Results:** A difference was highly positive association between knowledge, reported practice, & reported behavior (.674 & .398) respectively. In addition, the highest positively association with total self-reported practice and reported behavior with r (.608). **Conclusion:** Current study concluded that an educational program affecting positively on mother's knowledge and practices and behavior. **Recommendation:** Provide continuous health education to mothers and their children with PKU to improve their adherence to therapeutic regimen.

Keywords: phenylketonuria, mothers, practice, educational program.

Introduction

Phenylketonuria (PKU) is a disorder of amino acid metabolism caused by deficient phenylalanine hydroxylase (PAH) enzyme leads to accumulation of phenylalanine in the blood resulting profound and irreversible intellectual disability with cognitive and behavioral abnormalities if untreated (Lichter-Konecki and Vockley, 2019).

The incidence of (PKU) varies widely in different human population. United States are affected at a rate of 1 in 10,000. Turkey has the highest documented rate in the world, with 1 in 2,600 births, incidence of 1 in 4,500 in the Irish, while countries such as Finland and Japan have extremely low rates with less than one case of Phenylketonuria in 100,000 births (Dalei and Adlakha, 2022).

Diagnosis is by detecting high phenylalanine levels and low tyrosine levels and

altered ratio of phenylalanine to tyrosine in the untreated state with normal BH4 cofactor. Treatment is lifelong dietary phenylalanine restriction. Prognosis is excellent with treatment (Singh et al., 2021).

Mothers play a pivotal role in the management of childhood PKU which needs a diverse range of complex skills and learning programs to deal with the children disabilities and to maintain a special rehabilitation and diet program (Abdelrahim, Ahmed and Mostafa (2013), Kenneson, Youngborg & Singh, (2020).

The nursing interventions for children with PKU are pivotal. In particular, nurses should inform family caregivers about strict adherence to dietary protocol. Children suffering from long-term conditions require continuing support and nursing care throughout their lives. The nursing care of the child with PKU primarily aimed to supporting the child

and their families for minimizing the undesirable effect of illness (Marcason, 2014).

PKU is a burden on our Egyptian families especially in Upper Egypt due to high rate of consanguinity. PKU is a potentially serious inherited disorder. Without early diagnosis and correct treatment, most PKU childrens will develop irreversible brain damage, neurological problems and behavioral abnormalities (Sadek et al., 2013).

Unawareness of mothers about Newborn Screening (NBS) program and the lack of knowledge about best methods of dealing with their children with PKU, lead to the major aggravation of the disease problem.

The responsibility of pediatric nurse as a health educator in finding out the most applicable method of teaching, enables the caregivers mainly mothers to overcome the knowledge and practice deficit related to their children health needs and health problems.

The aim of the study

This study aims to evaluate effect of educational program on mothers' knowledge, practices and behavior towards caring for their children having Phenylketonuria through:

- 1- Assessing the mothers' knowledge about PKU.
- 2- Assessing the mothers' self-reported practices and behavior towards caring for their children with PKU.
- 3- Designing and implementing of educational program for mothers practices toward caring for their children with PKU.
- 4- Evaluating the outcome of educational program to overcome the deficit of mothers' knowledge and reported practices and behavior.

Research hypothesis

This study hypothesized that implementing of educational program will have a positive effect on knowledge and practices

and behavior of mothers towards caring for their children with PKU.

Subject and Methods

Research Design

Pre/post quasi experimental design was utilized to conduct the study.

Research Setting

This study conducted at the Metabolic Clinic, Department of Medical Genetics, Faculty of Medicine, Ain Shams University.

Subjects

A purposive sample of 128 mothers was selected according to certain inclusion criteria and exclusion criteria. The sample size was determined statistically by power analysis considering the total number of PKU children.

Inclusion Criteria

The study subject was selected according to the following inclusion criteria:

1. Mothers having children suffering from classical PKU.
2. Mothers having children aged from birth until 18 years of age and from both genders.

Exclusion criteria

Children having variant PKU & illiterate mothers.

Sample size

A purposive sample was utilized to conduct this study. The pre-post analysis will include numeric scores that can be transformed into categorical variables. A sample size of at least 128 mothers accompany their children having PKU will be satisfactory to detect an effect size of at least 0.25 (small to moderate effect size) using paired t-test at level of significance of 0.05 and power of 0.80. This sample size will be satisfactory to estimate the proportion that had improved knowledge/ reported practice of 50% with 95% confidence interval of 20% (40% to 60%).

Tools for data collection

Data was collected using the Pre/post designed interviewing questionnaire

It was written in a simple Arabic language based on updated references by the researcher to meet mothers' needs and suit their level of understanding and reviewed by supervisors. It consists of 4 parts:

Part 1: It concerned with Personal characteristics of mothers as: Age, level of education, residence, income.

Part 2: It concerned with mothers' knowledge about PKU was adapted from (van Spronsen et al., 2017) including questions regarding: definition, causes, symptoms and signs, diagnosis, prevention of potential complications, and different methods of treatment (dietary restriction, special medical formula and medical treatment).

Part 3: It concerned with Mothers' reported practice was developed after reviewing the current literature (National PKU Alliance, 2011) distributed along 8 domains measurement regarding measurement of 1) medical diet (7 items), 2) follow up of baby weight at home (5 items), 3) follow up of child weight at home (5 items), 4) follow up of length at home and 5) follow up of height at home (10 items), 6) follow up of (BMI) (4 items), 7) dental care (7 items), and 8) physical activity (5 items).

Part 4: It concerned with mothers' behavior was adapted from according to (Harrison and Oakland, 2015). It was distributed along 5 domains related to 1) communication (23 items), 2) community use (20 items), 3) leisure (20 items), 4) work (21 items) and 5) social skills (26 items).

Validity

Tools of this study were judged by a panel of 5 professor's expertise and they were professor of medical genetics & pediatric nursing. The necessary modifications were done according to experts' opinion to ensure validity of the content.

Reliability

The reliability for tool was 0.85 using Alpha Cronbach.

Ethical considerations

The study revised by the ethical committee at faculty of nursing Ain Shams University November 2020. Research approval for this study was obtained from the Dean of Faculty of Nursing, Ain Shams University to the Medical Genetic Department, Head of the previously mentioned setting. Oral consent was obtained from each mother prior to participate in the study. Participants in the study were assured that all information obtained would be kept confidential and that they had the right to withdraw from the study at any time.

Pilot study

A pilot study was carried out during November 2020 involved 10% (13 of mothers having children with PKU) of the total study sample. The result of the data obtained from the pilot study helped in removing of some repeated questions related to knowledge to avoid duplication of questions and then all children involved in the pilot study were included of the study sample.

Study framework

The actual field work was carried out over a period of 9 months from beginning of May 2021 up to the end of January 2022. The researcher was available in the study setting 4 hours for 2 days/week by rotation according to the clinic time to collect data.

The study was conducted in the following four phases: **A pre-program assessment test**, this phase aimed at assessing mothers' knowledge, reported practice, and reported behavior of PKU was conducted through interviews using the aforementioned questionnaire sheet. **Developing an educational program** the goals, priorities, and expected outcomes from the assessment phase

were formulated to meet mothers' needs in order to embed the self-learning package.

During the implementation phase, the studied mothers attended three consecutive educational program sessions, with the duration of each session ranging from 30 to 40 min. using effective media of conveying information as, laptop, posters, and power point presentation. The first session was regarding knowledge, including the definition of PKU, causes, signs and symptoms, diagnosis, prevention of potential complications, and different methods of treatment.

The second session included mothers practices related to formula preparations, follow up for growth as: (weight, height or length, Body Mass Index "BMI"), dental care, physical activity.

The third session focused on mothers' reported behavior towards their children having the disease related to self-care, self-direction, home living, functional academic, health and safety, communication, leisure, and social skills. In each session, small groups (ten mothers each) were led through small group discussions and assignments directed by the researcher.

In the evaluation phase (post-test), the same pretest questionnaire was used to assess the effect of self-learning package on mother's knowledge, reported practices' and reported attitude towards caring for their children having phenylketonuria. The questionnaire was administered immediately after the end of the training program.

Statistical Design

Data entry and statistical analysis were done using SPSS 23.0 statistical software package. Data were presented using descriptive statistics in the form of frequencies and percentages. Means, median and standard deviations for quantitative variables. T-test is a type of inferential statistic used to determine if there is a significant difference between the means of two groups. Qualitative categorical

variables were compared using chi-square test. Spearman rank correlation was used for assessment of the inter-relationships among quantitative variables and ranked ones. Statistical significance was considered at p-value <0.05.

Results:

Table (1): Reveals that more than half (55.5 %) of studied mothers' age were 20-<30 years old with mean score 23.7 ± 8.2 . As regard an educational level, less than two fifths of the studied mothers had intermediate education. More than two thirds of studied mothers were housewife. Moreover less than two thirds of them had insufficient income and coming from rural areas.

Table (2): Knowledge about PKU among studied mothers pre -post program, it was observed that about less two thirds had incorrect knowledge about definition preprogram, Regarding symptoms of the disease, less than one fifth of them had correct knowledge while posttest become 32.8% of them had correct knowledge. While majority of the studied mother had incorrect knowledge about types of foods that are not allowed at all, while post program, more than half (58.6 %) of studied mother had incorrect knowledge.

Nearly three quarters (73.4%) of the studied mother had incorrect knowledge on related to kind of exercise is appropriate for their child, while post program, 55.5 % of them had incorrect knowledge. Concerning mothers role toward their child performing exercise revealed that less than one third (29.7%) of the studied mothers had correct knowledge and become 46.1 % of them post program. Lastly, there was a high statistically significant difference in the level of knowledge among studied mothers about PKU among pre - post program as shown in Table (2).

Table (3): This table illustrates that more than three quarters 75.8% of the studied mother had unsatisfactory knowledge level at preprogram, while post program, 48.4% of them

had unsatisfactory knowledge level at p value (0.000**). Meanwhile, there was a high statistically significant difference in the total knowledge scores among studied mothers about PKU revealed to pre – post program.

Table (4): Reveals that there is a significance difference regarding self-reported mothers' practice level caring their children with PKU between pre and post program. 92.9% of mothers reported good practice regarding follow up of baby length post program while only 42.8% mother reported good practice preprogram. 46.8% of mothers reported good practice regarding Follow up of Body Mass Index post program while only 1.5% mother reported good practice pretest. 76.6% of mothers reported good practice regarding dental care post program while only 5.4% mother reported good practice preprogram. Regarding physical activity 60.1% of mothers reported good practice post program while only 21.9% mother reported good practice preprogram

Figure (1): This figure reveals that more than half of the studied mothers had poor practice level at preprogram, while only (3.1%) of them had poor practice level at post program. It is

observed that, there was a high statistically significant difference in the total level of practice among studied mothers of the study at different periods of measurements.

Table (5): Demonstrates that there is a significance difference regarding self-reported mothers' behavior level caring their children with PKU between pre and post program. 57.1% of mothers reported good behavior regarding time leisure post program while only 11.7% mother reported good behavior preprogram. 39.8% of mothers reported good behavior regarding communication post program while only 26.6% mother reported good communication preprogram. 38.3% of mothers reported good behavior regarding home living post program while only 17.2% mother reported good behavior preprogram. 36.7% of mothers reported good behavior regarding functional academic post program while only 30.5% mother reported good behavior preprogram.

Figure (2): This figure presents more than three fifth of the studied mother had poor behavior level preprogram, while more than one third of them had poor level of behavior post program.

Table (1): Personal characteristics of the studied mothers (N=128).

Personal characteristics	(n=128)	
	Frequency	Percent
Age group: (year)		
< 20	21	16.3
20 to < 30	71	55.5
30 to < 40	24	18.8
≥ 40	12	9.4
Mean ± SD		23.7 ± 8.2
(range)		(18 – 45)
Education:		
Basic education	46	36
Intermediate education	49	38.2
High education	18	14.1
Post graduate	15	11.7
Mother job:		
Working	42	32.8
Not working\house wife	86	67.2
Monthly Income:		
Insufficient	82	64.1
Sufficient	46	35.9
Residence:		
Rural	80	62.5
Urban	48	37.5

Table (2): Distribution of studied mothers' knowledge related to PKU pre/post program (n=128).

Knowledge about PKU	Pretest				posttest				X ²	(p-value)
	Correct		Incorrect		Correct		Incorrect			
	No	%	No	%	No	%	No	%		
Definition	47	36.7	81	63.3	94	73.4	34	26.6	26.86	.000**
Causes of the disease	26	20.3	102	79.7	60	46.9	68	53.1	36.97	.000**
Symptoms of the disease	23	18.0	105	82.0	42	32.8	86	67.2	57.41	.000**
Signs of the disease	26	20.3	102	79.7	58	45.3	70	54.7	39.37	.000*
Investigation	37	28.9	91	71.1	53	41.4	75	58.6	73.65	.000**
Times of phenyl alanine checking	34	26.6	94	73.4	57	44.5	71	55.5	57.66	.000**
Mothers role during checking phenylalanine level	38	29.7	90	70.3	59	46.1	69	53.9	63.21	.000**
Treatment methods for this disease	20	15.6	108	84.4	48	37.5	80	62.5	39.51	.000**
Types of foods that are not allowed at all	21	16.4	107	84.6	53	41.4	75	58.6	29.4	.000**
Types of foods that are allowed, but with an accounting	37	28.9	91	71.1	53	41.4	75	58.6	21.6	.000**
Kind of exercise is appropriate for child with PKU.	34	26.6	94	73.4	57	44.5	71	55.5	47.0	.000**
Mothers' role during child's exercise.	38	29.7	90	70.3	59	46.1	69	53.9	41.02	.000**
Impact of the disease on the child in social events and celebrations.	20	15.6	108	84.4	48	37.5	80	62.5	38.02	.000**

*: Significant **: Statistically Highly significant

Table (3): Distribution of total mothers' knowledge regarding PKU pre/ post program throughout the study phases (n=128).

Total knowledge about SLP	Pre (n=128)		Post (n=128)		χ^2	(p-value)
	No	%	No	%		
Total Knowledge score Satisfactory Knowledge	31	24.2	66	51.6	11.580	.000**
Unsatisfactory Knowledge	97	75.8	62	48.4		
Total mean score Mean \pm SD	6.11\pm4.33		14.81\pm1.68		t-test 10.523	.000**

** : Highly statistically significant

Table (4): Distribution of mothers' self-reported practice pre –post program (n=128).

Items		Pre(n=128)		Post(n=128)		χ^2	(p-value)	
		No	%	No	%			
1-Measurement of medical diet	Poor	70	54.7	13	10.2	15.098	<0.01**	
	Average	48	37.5	73	57			
	Good	10	7.8	42	32.8			
2- Follow up of weight	Poor	0	0	0	0	17.008	<0.01**	
	A- Baby weight (n=42)	Average	9	21.4	2			4.8
	Good	33	78.6	40	95.2			
B- Child weight (n=86)	Poor	10	11.6	3	3.5	14.760	<0.01**	
	Average	31	24.2	16	18.6			
	Good	45	35.2	67	77.9			
3- Follow up of height	Poor	12	28.6	0	0	16.003	<0.01**	
	A- Baby length (n=42)	Average	12	28.6	3			7.1
	Good	18	42.8	39	92.9			
B- Child height (n=86)	Poor	32	37.2	2	2.3	15.002	<0.01**	
	Average	24	27.9	13	15.1			
	Good	30	34.9	71	82.6			
4- Follow up of Body Mass Index	Poor	88	68.8	18	14.1	13.661	<0.01**	
	Average	38	29.7	50	39.1			
	Good	2	1.5	60	46.8			
5- Dental care	Poor	49	38.3	4	3.1	16.021	<0.01**	
	Average	72	56.3	26	20.3			
	Good	7	5.4	98	76.6			
6- Physical activity	Poor	48	37.5	13	10.2	18.055	<0.01**	
	Average	52	40.6	38	29.7			
	Good	28	21.9	77	60.1			

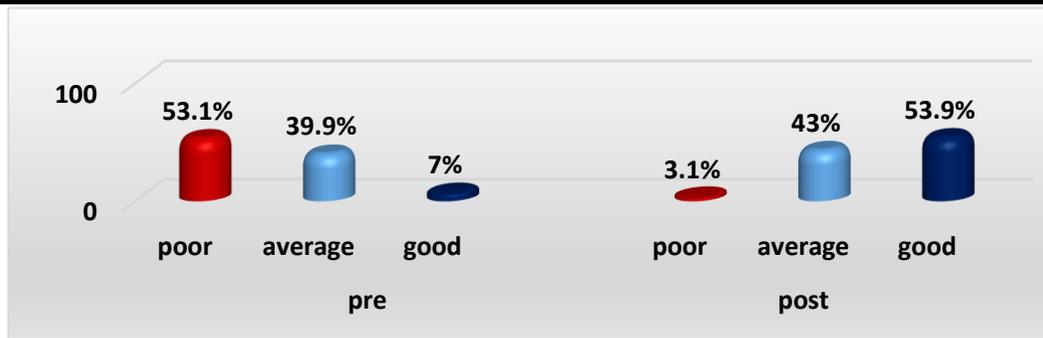
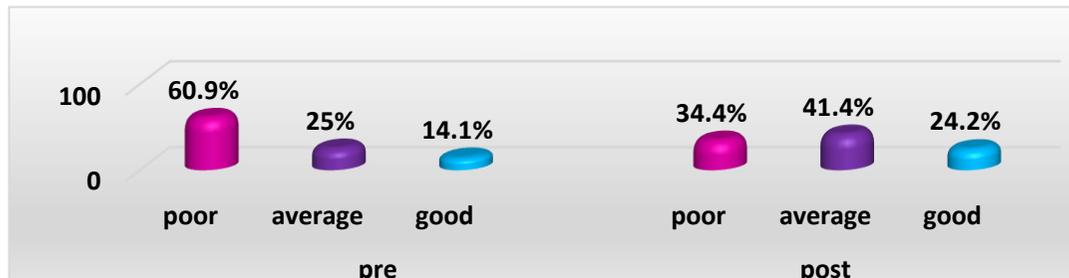
**Figure (1):** Distribution of total Self-reported mothers' practice pre –post program (n=128).

Table (5): The mothers reported behavior pre –post program.

Items		Pre(n=128)		Post (n=128)		χ^2	(p-value)
		No	%	No	%		
1-Practical	Poor	60	46.9	50	39.1	12.004	<0.01**
	Average	48	37.5	36	28.1		
	Good	20	15.6	42	32.8		
Home living	Poor	56	43.7	30	23.4	15.661	<0.01**
	Average	50	39.1	49	38.3		
	Good	22	17.2	49	38.3		
Health and safety	Poor	71	55.5	25	19.5	9.576	<0.01**
	Average	30	23.4	60	46.9		
	Good	27	21.1	43	33.6		
2- Conceptual	Poor	63	49.2	29	22.7	13.076	<0.01**
	Average	31	24.2	48	37.5		
	Good	34	26.6	51	39.8		
Functional academic	Poor	59	46.1	23	17.9	12.611	<0.01**
	Average	30	23.4	58	45.3		
	Good	39	30.5	47	36.7		
Self-direction	Poor	69	53.9	31	24.2	14.521	<0.01**
	Average	55	42.9	68	53.1		
	Good	4	3.2	29	22.7		
3- Social	Poor	36	28.1	18	14.1	16.913	<0.01**
	Average	72	56.2	65	50.8		
	Good	20	15.6	45	35.1		
Leisure	Poor	75	58.6	20	15.6	15.066	<0.01**
	Average	38	29.7	35	27.3		
	Good	15	11.7	73	57.1		

**Figure (2):** Distribution of mothers' reported behavior level pre –post program.

Discussion

Phenylketonuria is a rare inborn error of metabolism which needs caregiver's daily effort to maintain a special diet program, rehabilitation and to deal with the child's disabilities. Maintaining longtime nutrition regimens and applying complementary diets are the main therapeutic tasks in treating children with PKU which require perfect collaboration of the caregivers (*Jurecki et al., 2017*),

Family caregivers play a pivotal role in the management of childhood Phenylketonuria. Achieving quality of care for children with chronic or disabling conditions depends on embracing a family-centered perspective, so it is important to recognize, respect, assess and address family caregiver's knowledge, practice and needs. Establishing caregiver assessment as a basic component of practice across care settings, with the goal of strengthening those (*Gharai et al., (2011), Huckleberry et al., (2009)*)

In the present study, regarding the mothers' age it was revealed that more than half of studied mothers' age were 20 to less than 30 years old with mean \pm SD 23.7 ± 8.2 . This is anticipated because it is the normal age of childbearing.

Concerning educational level, the present study showed that less than two fifths of the studied mothers had intermediate education, this finding supported by Etemad et al., (2020) showed that nearly one third of the studied mothers had intermediate education.

The present study illustrated that more than three quarters of the studied mother had unsatisfactory knowledge level regarding phenylketonuria at pretest, while posttest, less than half of them had unsatisfactory knowledge level about PKU. Meanwhile, there was a high statistically significant difference in the total knowledge scores among studied mothers about PKU revealed to pre –posttest. This result matched with Abd-Elkodoos et al., (2018) who found that, 14.0% of the mother had good level of knowledge pre implementation of program compared to 74% of them post of program implementation.

Regarding to total Self-reported mothers' practice pre –posttest, the present study noticed that more than half of the studied mothers had poor practice level at pretest, while only (3.1%) of them had poor practice level at posttest. Additionally, there was a high statistically significant difference in the total level of practice of among studied mothers of the study at pre-posttest. These findings in same line with a study by Rahgoi et al., (2019) reported that there was a high statistically significant difference in the total level of practice of among studied mothers of the study at pre-posttest.

The current study showed that more than three fifth of the studied mother had poor behavior level towards caring their PKU children pretest, while more than one third of them had poor level of behavior posttest. Moreover, there was a high statistically significant pre, posttest. This finding consistent

with result done by Zamani et al., (2021) revealed that identifying barriers, self-monitoring, providing opportunities for social comparison and environmental changes. SLP had a more successful effect in improve mothers' practice.

The present result indicated that the there was a highly positive association between knowledge of studied mother and self-reported practice, reported behavior, the highest positive. In addition, the highest positively association with total self-reported practice and reported behavior with $r (.608)$. This finding might be due to that if there is adequate knowledge already there will be satisfactory practice and behavior.

This result agreement with study by Abd-Elkodoos et al., (2018).

who conducted study presented that the there was a highly positive association between knowledge of studied caregiver and self-reported practice This finding might be due to the mothers have a satisfactory knowledge about their children condition; this will improve their practices regarding care of their children with PKU and enhance their caring roles.

Conclusion:

Based on findings of the current study concluded that an educational program affecting positively on mother's knowledge and practices and behavior.

Recommendation:

In view of the study findings, the following recommendations are suggested: Continuous health education to mothers and their children with PKU to improve their adherence to therapeutic regimen. Repetitions of this research on large sample and in various places in Egypt and makes the mothers of children with PKU the focus of the health care team to improve their practices & improve their children lifestyle.

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Conflict of interest:

None

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