

FAMILY CAREGIVERS' KNOWLEDGE AND PRACTICES AMONG CHILDREN WITH PHENYLKETONURIA: A SUGGESTS NURSING CARE PROTOCOL

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ABSTRACT

Phenylketonuria (PKU) is a chronic disorder which needs family caregivers' daily effort to deal with the child's disabilities and to maintain a special rehabilitation and diet program. Family caregivers play a pivotal role in the management of childhood Phenylketonuria. The aim of the study was to suggest a protocol of care for family caregivers of children with phenylketonuria based on their needs assessment. A Methodological research design was utilized to fit the aim of the study. The study was conducted at the children clinic of metabolic disorder of the Social Preventive Medicine Center, affiliated to Faculty of Medicine, Cairo University. A purposive sample of 50 family caregivers and their children with PKU were included in the study. Data was collected using the following tools: I-Family caregivers structured questionnaire, II-Family caregiver's knowledge assessment questionnaire (Needs assessment), III- Observational checklist of family caregivers and IV- Needs assessment structured questionnaire for PKU children. The study revealed that 52.0% of the PKU children were females with a mean age of 7.8 ± 4.8 , all family caregivers were females with a mean age of 37.7 ± 8.1 and 64.0% of them had poor knowledge. The majority of the family caregivers had an unsatisfactory level of practice. A highly statistically significant positive correlation was found between the family caregivers' total knowledge scores and their total practices scores which means that the family caregivers' knowledge had an effect on their practices. This study proved that the nursing care protocol significantly improved the family caregivers' knowledge and practices related to the care of their children with PKU. The study recommended the publication and dissemination of the nursing care protocol in all maternal and child health care centers for families having children with PKU to provide knowledge and skills help them in caring for their PKU children at home and to raise their awareness about the disease management to prevent its complications

KEYWORDS: Family Caregivers, Phenylketonuria Children & Nursing Care Protocol

INTRODUCTION

Children of today and tomorrow are central to sustainable development and the future of our planet and all its populations. Healthy children are vital resources to ensure the future well-being of nations because they are the parents, workers, leaders and decision-makers of tomorrow, and their health depends on today's decisions and actions,

also their future lies in the hands of those people responsible for their well-being [1]. All parents want their children to grow up to live long, healthy lives, yet unfortunately, not all children have the same opportunity to be healthy. Factors such as where children live, how much education their family caregivers have and their race and ethnicity can make a real difference in their health as children and as adults [2]. Infants with untreated PKU seem to be normal for many months. However, without treatment, phenylalanine accumulates resulting in a mental deficiency, microcephaly, seizures, hyperactivity and purposeless movements, and eczema. [3]. As the PKU children are unable to efficiently metabolize phenylalanine, a component of protein found in normal diets, when PKU is untreated or treated late, the elevated blood Phe in PKU children can lead to severe mental retardation or reduced IQ, seizures and tremors, difficulties in executive functions, psychological and behavioral issues, social difficulties, impaired growth, irritability, and eczema [4]. However, with early and continuing dietary management, children with PKU have normal physical and cognitive development [5]. PKU is one of the first genetic disorders in which early treatment intervention has prevented clinical symptoms, increased the productivity of affected children, and decreased societal burden [6]. Treatment for PKU is focused on maintaining a safe level of Phe concentration in the blood, promoting normal growth and health through life and preventing intellectual disability [7]. Family caregivers play a pivotal role in the management of childhood Phenylketonuria [8]. PKU is a chronic disorder which needs family caregivers' daily effort to deal with the child's disabilities and to maintain a special rehabilitation and diet program [9]. Achieving quality of care for children with chronic or disabling conditions depends on embracing a family-centered perspective. Because family caregivers are a core part of health care and long-term care, it is important to recognize, respect, assess and address family caregivers' knowledge, practice and needs. Establishing caregiver assessment as a basic component of practice across care settings, with formal recognition of caregiving families and the goal of strengthening them; calls for a fundamental change of thinking in policy and practice [10]. Designing a Protocol for family caregivers of PKU children is framework outlining the care that will be provided to children in a home and it describes why, where, when and by whom the care is given. The Modernisation Agency has suggested that protocol of care provides clear statements and standards for the delivery of care locally. Care protocol is intended to provide information, based on an appraisal of the current best evidence of clinical and cost-effectiveness, regarding interventions for PKU children [11].

METHODS

Aim of the Study

The aim of the current study was to suggest nursing care protocol for family caregivers of children with phenylketonuria based on their needs assessment. This aim was covered by six main objectives:

- Assess family caregivers' knowledge and practice regarding care of Phenylketonuric children
- Assess the bio-psycho-social needs of Phenylketonuric children.
- Suggest a nursing care protocol based on needs assessment for both family caregiver's and their children.
- Implement the suggested protocol of care.
- Evaluate the outcome of the suggested protocol of care for both family caregiver's and their children.
- Validate & Reliable of the suggested nursing protocol of care.

Operational Definition

Family Caregivers

A family caregiver is the person who provides direct care for a child with PKU in the family.

Need Assessments

Need assessment is a systematic set of procedures that are used to determine needs, in a present state (what is) & (what should be) for future action.

Research Design, Participants & Setting

The Methodological design was utilized in the current study. The study was conducted at the children clinic of metabolic disorder, at Center for Social and Preventive Medicine affiliated to Faculty of Medicine, Cairo University and Ministry of health. A purposive sample of 50 family caregivers who provide direct care for a child with PKU in the family was utilized in this study.

Ethical Considerations

An approval of the ethical committee of nursing faculty, Cairo University was obtained upon the feasibility of the study tools. An official permission to conduct the proposed study was obtained from the director of the Social Preventive Medicine Center at Abo El-Reesh. Participation in the study is voluntary and based on the studied sample ability to commit to the current study; where written informed consent was signed by the participants who accept to be included in the study after reading all its details; the ethical issues considerations include explaining the purpose and nature of the study, stating the possibility to withdraw at any time, confidentiality of the information will be assured through coding the data, where it will not be accessed by any other part without taking permission of the participants and participation is with no risk.

Data Collection Tools

Family caregivers structured questionnaire: It was developed by the researcher, it includes two parts: A- Demographic characteristics of the family caregivers, it covers 26 closed-ended questions which includes age, gender, place of residence, relation degree, education, employment, marital status, health status, medical history etc... According to Orem's theory of self-care, these characteristics are the basic conditioning factors which affect the ability to perform the self-care. B- Socio- economic data of the family caregivers, it is composed of: Socio-economic status scale (SES) which developed to assess the socio-economic status of the Egyptian family. It was created by (El-Gilany, El-Wehady & El-Wasify, 2012).It was tested for validity & reliability. It includes 7 domains: (1st domain assessed different levels of education and cultural, 2nd domains included questions related to different types of occupation, 3rd domains related to family numbers, while 4th domains are related to family possessions, 5th domains covers economic status and source of income, 6th domains assessed Home sanitation, and 7th assessed Healthcare domains). II- Family caregiver's knowledge assessment questionnaire (Needs assessment): it was developed by the researcher to assess the family caregivers' knowledge regarding care of their PKU children, it includes 93 multiple choice questions in six sections: Background knowledge regarding PKU disease (25 questions), opinions regarding the services provided (15 questions), Child's health status (5 questions), Breast feeding & artificial feeding (17 questions),

Child's nutrition (26 questions), Child's growth (3 questions), and Social support (2 questions). The total scores of the knowledge was (186 marks) categorized as: poor level of knowledge (score < 93 marks), fair level of knowledge (score ranged from 93-139.5, and a good level of knowledge (score ranged from 140-186). III-Observational checklist of family caregivers: It was developed by the researcher to assess the practices of family caregivers regarding care of their PKU children. It includes 40 statements in six sections: Formula preparation (7 statements), feeding process (14 statements), dietary record (1 statement), bathing (6 statements), Hair and Nail care (4 statements), hand wash (3 statements), Dental care (4 statements), and Bathroom use (4 statements). Answers of the checklist were either done or not done, with the score of one mark for done and zero for not doing. The total scores for the observational checklist were 40 marks. Family caregivers' practices were classified into either satisfactory done (50% and more) or not done (less than 50%). IV- Needs assessment structured questionnaire for PKU children: It was developed by the researcher, it includes two parts: A- Personal characteristics of the PKU children, it covers 10 questions which include: age, gender, weight, height/length, education, child's age at detection time of PKU, Phe level (the initial test), Phe level (the current test), number of brothers & sisters and child's ranking between their siblings. Ranking of questions started from one to ten. According to Orem's theory of self-care, these characteristics are the basic conditioning factors which affect the ability to perform the self-care. B- Assessment of bio-psychosocial needs of PKU children. It covers 98 questions in two main sections, physical needs, and psychosocial needs. The first section covers 74 questions about physical needs assessment that includes questions about: concept of nutrition (11 questions), hygiene care (29 questions), elimination/excretion (2 questions), sleep & rest (9 questions), clothing (2 questions), daily life activities (3 questions), education (6 questions), health (7 questions), play and activities (5 questions). The second section covers 24 questions concerned with psychosocial needs assessment which includes questions about: love & intimacy (3 statements), self-esteem (9 statements), social estimation (5 statements), and emotional safety (7 statements). Answers to this part ranged between yes, sometimes and no with the scoring of two, one, and zero respectively. The total scores obtained on bio-psychosocial needs of PKU children were 196 classified as: scores from 0: 97 referred to low expressed needs, scores from 98: 147 referred to average expressed needs, scores from 148: 196 referred to high expressed needs.

Data Collection Procedure

The study was conducted in sixteen months from July 2015 until November 2016. It was carried out in six phases; administrative phase, needs assessment phase, planning phase, implementation phase, evaluation phase, validity and reliability phase.

A- Administrative phase: An official permission was obtained from the director of Center for Social and Preventive Medicine which is affiliated with Faculty of Medicine, Cairo University. Also, an official permission was obtained from the director of clinics, then permission from the director of metabolic disorder clinic. As well, written and oral consent was obtained from all the family caregivers who participated in the study. Written consent was obtained from the representative of the family caregiver In case of non-reading and writing

B- Needs assessment phase: Data was collected by the researcher using four tools. Two to three family caregivers were interviewed one day/week from 8.00 am to 2.00 pm. This day was specific for PKU children attending with their

family caregivers weekly in the metabolic disorder clinic. The time spent to fill each questionnaire sheet ranged from 30 to 35 minutes. Regarding observational checklist, family caregivers were observed while they were performing care for their PKU children. Time spent to fill each observational checklist ranged from 50 to 60 minutes. The total sample after six months (from July to December 2015) was 50 family caregivers with their PKU children. The data was submitted for the second time to the ethical committee for evaluation and final approval was obtained on 8-1-2017

C-Planning and designing phase: Based on the actual result that obtained from needs assessment using the interviewing questionnaire, observational checklist as well as literature review of the current and past local and international related books, magazines and periodicals which aimed to satisfy the studied family caregivers deficit knowledge and practice regarding to care of their children with PKU, the researcher developed a nursing care protocol for family caregivers with PKU children. It was designed in English and translated into Arabic. The content of the designed protocol included goal, objectives, simple and clear information about PKU definition, its causes, types, signs, symptoms, dietary management, importance of life long complying with dietary formula, food recording, monitoring blood Phe level and how to apply at home, importance of oral care for PKU child and how to maintain it, educating PKU child to be independent to satisfy his/her basic needs, complications of PKU, its impact on the child, guide lines for implementing care for PKU children at their homes, and the role of the family caregivers. This phase was over a period from January to March 2016.

D-Validity & Reliability phase: The suggested nursing care protocol for the family caregiver with PKU children has been subjected to validity & reliability test before proceeding to the pilot phase. Content validity was tested by jury of (11) experts: from faculty of nursing (3) professors, pediatric medicine (3) professors, molecular medical genetics (3), and food science (2) professors at Cairo and Ain-shams to confirm that this protocol focuses on the needs of the PKU children and their family caregivers and to check the adequacy of items that cover the domain under suggestion. Based on experts' recommendations needed modification was done and the nursing care protocol for family caregiver with PKU children were constructed. This phase lasted from April till May 2016. The researcher designed an Opinionnaire sheet to test the content validity of the constructed nursing care protocol for a family caregiver with PKU children to fill by the jury group; it involved two parts:

1-The opinions of the experts for each item which recorded in two points scale; relevant or irrelevant.

2- General or overall opinion about the form of the developed nursing care protocol: they asked to read the protocol and evaluate the content in terms of whether it appears to cover all dimensions of care for PKU children, clear concise and comprehensive. Express their opinions and comments on the developed nursing care protocol for family caregiver with PKU children and provide any suggestions for any adding or omitting of items. After completion of jury opinions, Opinionnaire sheets were reviewed and analyzed.

E- Implementation phase: The suggested nursing care protocol for a family caregivers with PKU children was implemented on forty family caregivers of the previous sample, this was the sample available during application of the protocol, but the printed version of the protocol was distributed to all family caregivers with PKU child who attending the metabolic clinic later on. This phase supported by a detailed training for the family caregivers who attend implementation of the protocol. The protocol was supported by written and oral instructions knowledge in addition to demonstration and

re-demonstration related to skills.

- The nursing protocol of care was conducted in the form of 8 sessions/ month).
- Family caregivers and their PKU children were divided into subgroups. Maximum number of family caregivers and their PKU children was ten in the session.
- Each group took 2 sessions/week/one month, in a form of knowledge and practical sessions. Each session will be disseminated to family caregivers of children with PKU in a class at the Social Preventive Medicine Center for (30-45) minutes. Thirty minutes in a form of lecture or demonstration, in addition to (15) minutes for open discussion to clarify and explain any misunderstanding.
- Data show posters booklets, video film, counseling method, demonstrations and re demonstrations were used.
- This phase conducted through June 2016.

Nursing care protocol content was prepared to be presented in Arabic and its content was built on the review of the related literature. This content was structured by the researcher based on Dorothea Orem's self-care deficit theory (2001). A full explanation of the study was explained orally and in written form. Every family caregiver took a copy of the Arabic Hand Book entitled: "family care for the PKU child: a simplified reference for each family"

F- Evaluation Phase: Evaluation of outcomes of the suggested nursing care protocol for the family caregiver with PKU children was carried out by the researcher immediately post protocol application and after three months to assess retention of acquiring knowledge and skills by using the same study tools. This phase lasted from August until September 2016.

Tools Validity and Reliability

Content and face validity was established by a panel of experts consisting of two professors of community health nursing, one professor of critical care and emergency nursing, one professor of pediatric health nursing, and one professor of maternal and newborn health nursing. Each of the experts on the panel was asked to examine the tool for content, clarity, wording, length, format, and overall appearance. Then required modifications were done accordingly.

Statistical Design

Upon completion of data collection, the data were coded, scored, tabulated, and analyzed by computer using the "statistical package for the social science" (SPSS windows) version 21. Frequency and percentage were used for numerical data as well as the mean \pm standard deviation, minimum and maximum. For finding the differences between normally distributed numeric data, t-test and Analysis of Variance One Way ANOVA test were used. For finding the differences between categorical data, nonparametric Chi-square (X²) test was used. Probability (p-value) less than 0.05 was considered significant and less than 0.001 considered as highly significant.

RESULTS AND DATA ANALYSIS

Part I: Description of Socio-Economic and Demographic Data of the Family

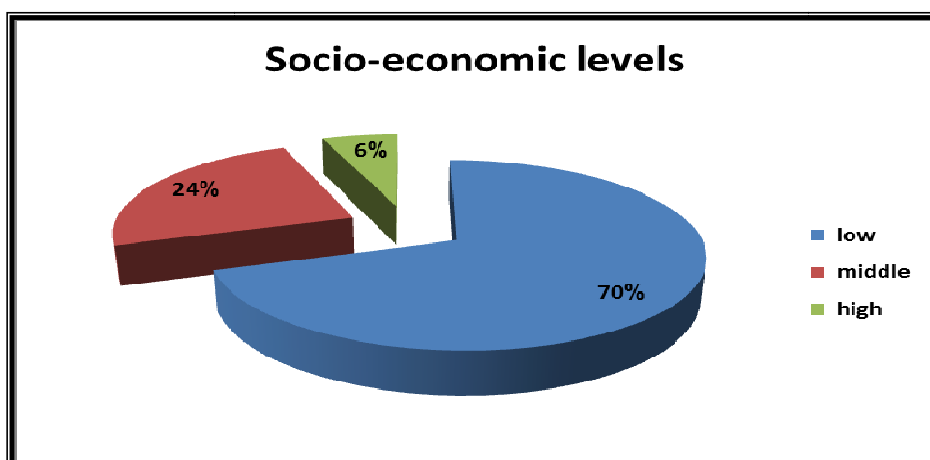


Figure 1: Percentage Distribution of Socio-Economic Levels Among Study Sample (n = 50)

Table 1: Frequency and Percentage Distribution of Socio-Demographic Characteristics of the Study Sample (n=50)

Socio-Demographic Characteristics	n=50	
	No	%
Caregiver's age:		
20-30	14	28.0
31-40	18	36.0
41-50	12	24.0
51-60	5	10.0
> 60	1	2
X ± SD	37.7± 11	
Sex:		
Female	50	100.0
Caregivers education:		
Doesn't read & write	8	16.0
Read & write	20	40.0
Secondary education	16	32.0
University education	6	12.0
Caregiver's marital status:		
Unmarried	1	2.0
Married	42	84.0
Widow	6	12.0
Divorced	1	2.0
Employment :		
Not work/ House wife	39	78.0
Professional work	4	8.0
Crafts	1	2.0
Free business	6	12
Relation to the child:		
Mother	47	94.0
Grandmother	2	4.0
Other relatives	1	2.0
Direct child care		
Father	1	2.0

Table 1 Contd.,		
Socio-Demographic Characteristics	n=50	
	No	%
Mother	47	94.0
Grandmother	1	2.0
Other relatives	1	2.0
Mother & father Consanguinity:		
No	20	40.0
Yes	30	60.0
Period of caring PKU child		
<1	1	2.0
1-5	17	34.0
6-10	18	36.0
11-15	10	20.0
>16	4	8.0
Other caregivers in the family:		
Yes	25	50.0
No	25	50.0
If yes, the other child caregivers is:		
Father	23	46.0
Grandmother	12	24.0
Sister	5	10.0
Brother's wife	1	2.0
Nursery	1	2.0
Intervening years between the birth of the PKU child & previous child:		
Less than one year	1	2.4
1 - 5 years	37	90.3
6 - 10 years	3	7.3
X ± SD	3.2±2.2	
Intervening years between the birth of the PKU child & next child:		
1 - 5 years	16	84.2
6 - 10 years	3	15.8
X ± SD	2.7±1.3	

*NB. The number in these variables didn't equal because many caregivers had only one or two child.

Table 2: Percentage Distribution of the Family Caregiver's of Children with PKU Regarding Pre-Marital Examination & Chronic Illness (n=50)

Variables	Total sample n=50	
	No	%
Pre-marital examination checkup:		
Yes	5	10.0
No	45	90.0
Benefits of this examination for whom:		
Wife	3	6.0
Children	9	18.0
All of the above	16	32.0
Not useful for anyone at all	22	44.0
You really benefited of this examination:		
No	50	100.0

Table 3: Frequency and Percentage Distribution of the Study Sample Regarding Family Past Medical History of PKU (n=50)

Variables	n=50	
	No	%
Chronic illnesses among family caregivers:		
Yes	29	58.0
No	21	42.0
Type of illness:		
Diabetic	15	30.0
Hypertension	28	36.0
Cardiac diseases	3	6.0
Renal diseases	5	10.0
Hepatic diseases	1	2.0
Lumbar disc	11	22.0
Previous PKU cases in the family:		
Yes	25	50.0
No	25	50.0
If yes, relationship to PKU child:		
Cousin	7	28.0
Sister / brother	14	56.0
Uncle	4	16.0
No. of PKU children in the family:		
One child	34	68.0
Two children	14	28.0
Three children	2	4.0

*NB. The number in these variables didn't equal because many caregivers had more than one chronic disease

Table 4: Frequency and Percentage Distribution of the Study Sample Regarding Family Past Medical History of PKU (n=50)

Variables	n=50	
	No	%
Other chronically ill children in the family		
Yes	6	12.0
No	44	88.0
If yes, disease type:		
Diabetes	5	80.0
Respiratory diseases	1	20.0
Does the PKU child suffer from other chronic diseases?		
Yes	21	42.0
No	29	58.0
If yes, disease type:		
Increased brain electricity	22	44.0
Autism	5	10.0
Cross-eyes	1	2.0
Brain atrophy	2	4.0
Convulsion	8	16.0
Epilepsy	1	2.0
Motor disability	1	2.0

*NB. The number in these variables didn't equal because many PKU children have more than one chronic disease.

Part II: Family Caregiver's Knowledge Assessment Questionnaire (Needs Assessment)

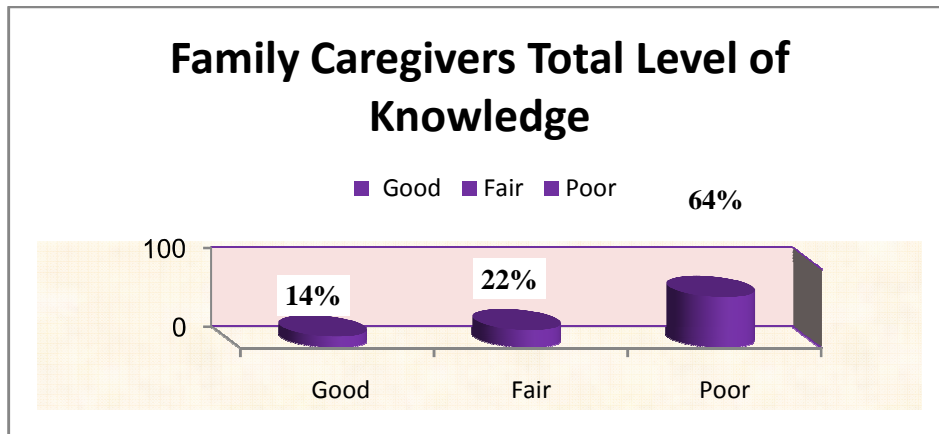


Figure 2: Family Caregivers' Total Level of Knowledge Regarding Care of Their PKU Children (n=50)

- Poor level of knowledge= <60% of total score
- Fair level of knowledge= 60% to 75% of total score
- Good knowledge= more than 75% of total knowledge

Part III: Observational Checklist for Family Caregiver's Practices Regarding Care of PKU Child

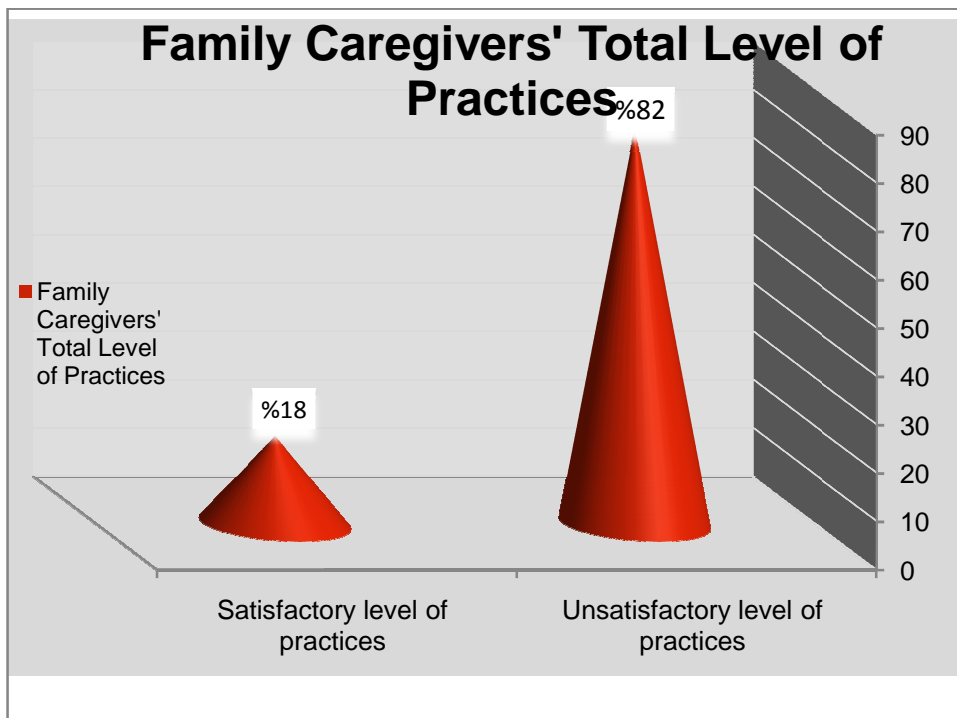


Figure 3: Family Caregivers' Total Level of practice regarding Care of Their PKU Children (n=50)

Table 5: Correlation between the Total Family Caregivers' Knowledge and Practices Scores & the Child Personal Characteristics (n=50)

Child's Personal Characteristics	Total Knowledge		Total Practice	
	r	p	r	p
Age	0.088	0.299	-0.262	0.002**
Sex	0.003	0.976	0.168	0.047*
Child's age at disease detection	-0.187	0.027*	-0.456	0.000**
Number of sisters & brothers	-0.209	0.021*	-0.101	0.272
Child rank	-0.107	0.208	-0.065	0.443
**. P value is highly significant at the level of ≤ 0.01				
*. Correlation is significant at the level of ≤ 0.05				

Table (6): Correlation between the Total Family Caregivers' Knowledge and Practices Scores & Their Socio-Demographic Characteristics & Health Status (n=50)

Caregivers Socio- Demographic Characteristics	Total Knowledge		Total Practice	
	r	p	r	p
Age	-0.074	0.386	-0.342	0.000**
Sex	-0.026	0.761	0.021	0.803
Education	0.457	0.000**	0.407	0.000**
Marital status	-0.078	0.362	-0.221	0.009**
Consanguinity	-0.245	0.027*	-0.128	0.257
Employment	-0.106	0.214	0.141	0.097
Monthly income	0.282	0.001**	0.054	0.540
Place of residence	0.400	0.000**	0.387	0.000**
Type of home(rent or estate)	-0.352	0.000**	-0.392	0.000**
Health status	0.083	0.331	-0.140	0.098
**. Correlation is highly significant at the level of ≤ 0.01				
*. Correlation is significant at the level of ≤ 0.05				

Table 7: Correlation between the Total Family Caregivers' Knowledge and Practices Scores and the Number of PKU Children in the Family, Background Knowledge of PKU & Period of Caring For the PKU Child (n=50)

Items	Total Knowledge		Total Practice	
	r	p	r	p
PKU children in family	-0.123	0.156	-0.206	0.017*
Background knowledge of PKU	0.502	0.000**	0.014	0.868
Period of caring to PKU child	0.116	0.172	0.166	0.050*
**. Correlation is highly significant at the level of ≤ 0.01				
*. Correlation is significant at the level of ≤ 0.05				

Table 8: Correlation between the Total Knowledge Scores of the Family Caregivers and Their Total Practices Score (n = 50)

Items	r	P
Total knowledge score	0.315	0.000**
Total practice score		

DISCUSSIONS

Results of the current study showed that more than two-thirds of the families their socioeconomic status was low, while almost one- fifth of them have middle socioeconomic status. This congruent with [12], who conducted his study on 186 family caregivers and their children with PKU, at a metabolic care center in Gaza Strip, at Al-Remal PHC in Gaza

city and found that, more than half of the study sample on the poverty line. These findings in contrast with [13] who conducted their study on 173 family caregivers and their children with PKU, at the Medical University, Bialystok, Poland, found that, over 70% of parents of PKU children described their family's economic conditions as very good or good, 20% as average and 1% as poor. This difference because the majority of family caregivers weren't working which leads to that the majority of them had insufficient family income and low socioeconomic level. A highly statistically significant positive correlation was found between the family's socioeconomic status and the total knowledge scores. With high socioeconomic status, there will be the availability of mobility/travel, which increases the chances of diagnosis, further investigations, and follow-up at the PKU clinic and thus increasing the chance of getting more knowledge about the disease and treatment by specialized doctors.

In the current study, it was found that the mean caregiver's age was 37.7 ± 8.1 years. A highly statistically significant negative correlation was found between the total practice scores of the family caregivers and their ages. This goes in the same line with [14] who conducted their study on 153 PKU children and their family caregivers in the Pediatric Genetics Clinic in Ain Shams University Children's Hospital and the Child Clinic of Phenylketonuria Metabolic Disorder in the Pediatric Hospital Campus of Cairo University and reported that, the mean age of the family caregivers was 35.3 ± 3.2 years and found a negative correlation between the total practice scores of the family caregivers and their ages. In the same context [15] conducted a study on 49 family caregivers of PKU children in the Child and Adolescent Consultation Liaison Psychiatry Clinic of children medical center in Tehran for a routine clinical follow up reported that, the mean age of family caregivers was 35.63 ± 8.82 years and also found a negative correlation between the total practice scores of the family caregivers and their ages. It seems that the greater the age of family caregivers, the less the level of practices and conversely. This is true because young family caregivers have the strength and the physical ability to look after their children.

In the present study, it was found that, all the family caregivers were females and most of them mothers of the PKU children; these results were supported by many studies as [12] who found that most family caregivers were mothers and reported that mothers of children are considered the most primary health care providers around the world. Also [16] who conducted a study on 140 family caregivers and their PKU children in the Children Clinic of Metabolic Disorder in Medicine Center in Cairo and found that, most of the family caregivers were the biological mothers of the child and reported that, the family caregivers usually are females because many of activities of daily living for children are most often done by females in families, especially by their mothers. [17] Conducted a study on family caregivers of children suffering from hemiplegic palsy at El-Menia pediatric hospital and reported that, all the family caregivers were mothers of the children; and [15] revealed that, almost two-thirds of the family caregivers were mothers of their children.

[18] Stated that, mothers of children are considered the most primary health care providers around the world. They take on enormous responsibilities in providing care and managing of their child's conditions outside the healthcare institutions. In Egyptian culture, mothers are also the main caregivers of their families with or without sick persons in their families. These previous explanations also in the same line with the known facts that females tend to give more emotional support than men.

Regarding the family caregiver's education, it was noticed that, more than half of the family caregivers were either illiterate or just can read and write and the minority of them had higher education. This finding was supported by [14] who reported that, more than one tenth of them were illiterate, and more than one-tenth of them can read and write, almost two fifth of them had intermediate education, while only less than one-fifth of them were having a university education. In the same context [17] found that, almost half of family caregivers were illiterate and the minority of them had higher education. In contrast to the study results [19] found that, most of the family caregivers completed high school, and approximately one-third completed college or graduate school. As well [20] whose study entitled "Implication of resolving the diagnosis of PKU for parents and children" revealed that half of the family caregivers had completed tertiary education. This could be related to the difference among cultures and that most of the females in Egypt are less educated than males and this is more common among rural than urban females as reported by [21]

There was a highly statistically significant positive correlation between educational status of the family caregivers and total knowledge scores and total practice scores as well, which was supported by several studies as [22]; [23]; [14], (2016) and [13] who reported that, family caregiver's knowledge scores increased with the family caregiver's educational levels which may improve the care for the PKU children while family caregiver's illiteracy was a negative factor in practice compliance. On the other hand, [24] conducted a study on 53 Swedish children with PKU with their family caregivers found no relation between the family caregiver's educational level and their level of practices. Educated family caregivers usually have more knowledge and practices compared with illiterate or less educated family caregivers.

As expected in the current study, a better adherence was observed with a high level of education of the family caregivers, but illiteracy of the caregivers decreased the rate of compliance with the rigid and long-term dietary treatment which could suggest the possibility of specific intervention directed towards less educated families. The difference between current studies and [24] could be related to the nutrition specialists in Sweden who train families and increase caregivers' knowledge independently from their education.

Regarding marital status, the majority of the family caregivers in the current study were married, while a minority were either widowed or divorced, which was consistent with many studies such as [19]; [12]; and [16] who reported that, most of the family caregivers were married and the minority were separated or divorced. There was a highly statistically significant negative correlation between the total practice scores of the family caregivers and their marital status. In spite of slight number of both divorced and widowed caregivers (7 out of 50) in this study, dietary adherence among these divorced and widowed women was significantly poorer than married ones, which is similar to results of [24] who found that divorced family caregivers had more difficulties in controlling their PKU children's care. This result denotes that, neglecting of the child's care could happen more common in divorced families because divorced mothers are very busy working and bringing money to support their children.

Regarding the family caregiver's occupation, the current study revealed that, the majority of the family caregivers were housewives and the minorities of them were working. This finding was supported by [14] who reported that the majority of caregivers weren't working. In contrast to the study results [13] who reported that most family caregivers were employed. This difference may be related to the difference in the traditions and believes among cultures where women

value their work very much in developed countries.

No statistically significant correlation was found between family caregiver's job and their total practice & knowledge scores. This was in agreement with the findings of a study done by [26] revealed that, family caregiver's knowledge scores were higher in currently employed caregivers. Another study done by [23] identified that, Phe dietary control provided by the employed family caregivers was significant than those provided by the unemployed. Also, a recent study was done by [13] who reported that, Phe dietary control and caregiver's level of practices for their PKU children was not affected by family caregiver's employment status. The difference in these results and the previous two studies could be related to the number of the family caregivers selected in the studies where the majority was unemployed and only 23 family caregivers were employed and worked females.

In relation to the caregiver's consanguinity, it was noticed in the current study that more than half of mothers and fathers of the PKU children were having the consanguineous relationship. There was a statistically significant negative correlation between the total knowledge scores of the family caregivers and their consanguinity. This result was supported by [27] who conducted a systematic review to examine twenty-nine studies reporting demographic and/or psychosocial influences on blood phenylalanine levels, with the aim to identify factors that were robustly linked with metabolic control and found that more than half of caregivers were having consanguineous relationship between father and mother of the affected child. This could be attributed to the scientific fact that PKU is a chronic genetic disorder and more common in consanguinity marriage.

Regarding the family caregivers' health status, more than half of the family caregivers were having chronic illnesses, while less than half did not have any chronic illnesses. No statistically significant correlation was found between family caregivers' illness and their total knowledge & practice scores. This could be attributed to that small number of ill family caregivers is still able to provide care to their PKU children.

The current study findings clarified that, more than one-quarter of the family caregivers were having more than one PKU child in their families. This agrees with the results revealed by [16]; and [14] who reported that who reported that almost one-quarter of the families had multiple children with PKU, also [13] who found that, more than one-third of the families had 3 or more PKU children. A statistical significant negative correlation was found between the total practice scores of the family caregivers and the number of PKU children in the family, while no significant statistical correlation was found between the number of PKU children in the family and the total knowledge scores. This result was in agreement with [23] who reported that, by increasing number of affected children in the family, there was poor Phe diet control and increase in blood phenylalanine concentrations. It seems that, as the number of affected children in the family increases there will be more pressure on the family resources either physical, psychological and/or financial resources. Also, family caregivers with one PKU child could easily seek new knowledge and try to improve their practices but family caregivers with more than one PKU child are very busy and have no time to seek more knowledge.

Concerning family caregiver's background knowledge about PKU, the majority of them reported having little information about PKU. They got their knowledge from nurses and doctors only, and the others got information from their relatives only. These results are in part different from [19] who described that, in most families caregivers were having multiple sources of access information regarding their PKU children, including health care providers, the internet,

support groups, and organizations associated with the condition. This difference may be related to the lower educational levels of the family caregivers plus their rural residency and lacking accurate information given by the health team in the metabolic clinic.

A highly statistically significance positive correlation was found between the family caregiver's background knowledge about PKU and their total knowledge scores, while no statistically significant correlation was detected with the total practices scores of the family caregivers. These results are logic as the background knowledge about PKU was obtained; the family caregiver's knowledge improved which reflect the need of those caregivers for effective nursing care protocol for PKU children.

More than one-third of the family caregivers were responsible for caring for children from 1 to less than 5 years. More than one third was responsible for children from 6 to 10 years, while just a minority were responsible for children less than one year. A statistically significance positive correlation was found between the total practice scores of the family caregivers and the period of caring for their children with PKU. No statistically significant correlation was found between the total knowledge scores and the period of caring for their children with PKU, which means that, the longer period the caregivers spend caring for their PKU children, the more their practices get improved. On the contrary, [13] found a negative correlation between duration of treatment and practices level on Phe diet control. This could be related to increasing the period of caregiving for the child plus the repetition of care would improve the caregiver's performance.

Concerning the definition of PKU, it was found that, more than three-quarters of the family caregivers could correctly defined PKU. This is in accordance with the study results [14] who revealed that the majority of the caregivers be defined PKU as a metabolic genetic disorder. [12] Reported that, the majority of family caregivers could correctly define the term PKU. Also, the most prevalent symptoms observed by more than two third of the family caregivers were a convulsion, while the minority reported the early signs and symptoms as follow: nervousness, irritability, musty odor in mouth and urine and change in color of hair and skin. These results are in part different from the study done by [13] where the majority reported that "irritability," "moodiness," or "whining" were the most prevailing symptoms. This difference may be due to lack of family caregiver's awareness about early symptoms of PKU and fluctuation in time of discovering the disease.

In the current study, data about the family caregiver's knowledge regarding phenylalanine test showed that, the majority of the family caregivers defined phenylalanine test (blood test only). Concerning the required level of blood phenylalanine, it was noticed that, more than two-thirds of the family caregivers reported did not know. On the contrary, the study done by [14] found that, the majority of family caregivers understood what was meant by a 'phenylalanine test and correctly defined phenylalanine, and identified correctly the recommended ranges of blood phenylalanine. The difference between this study and the previous could be related to unawareness of the family caregivers regarding phenylalanine test or may be due to their educational level.

Regarding the family caregiver's knowledge about complications of increasing the phenylalanine intake, more than two third of the family caregivers were aware of the most common complications such as mental retardation, delay in motor activities and delayed growth. This result agrees with [12], who reported that, the majority of the family

caregivers were aware of the adverse consequences associated with increasing the phenylalanine intake. This means that, in spite of their poor knowledge about early signs of PKU, still they are aware of the most common complications that could happen and this could be related to either that, they were faced with this complications or they knew that this disease could cause serious complications so they were interested in knowing them.

In relation to the family caregiver's knowledge about PKU treatment, it was observed that the majority of the family caregivers reported diet regimen as a treatment for PKU. This was in agreement with [24] who reported that, the majority of the family caregivers identified the type of treatment, and also by [28] stated that, family caregiver's knowledge concerning the diet was accurate for the majority of family caregivers. This result is true since diet is the only treatment for PKU, so the family caregivers knew this well, and also after the child is being diagnosed as having PKU, the family caregivers knew from the specialized physician that the diet was the only available type of treatment because medical treatment is not available in Egypt yet.

The results of the present study showed that 50 family caregivers and their children with PKU were included; the mean age of children with PKU was 7.8 ± 4.8 years. The same result was revealed from [28], [14]; who conducted their study on 153 PKU children and their family caregivers the Pediatric Genetics Clinic in Ain Shams University Children's Hospital and the Child Clinic of Phenylketonuria Metabolic Disorder in the Pediatric Hospital Campus of Cairo University and reported that, 8.1 ± 5.2 was the mean age of PKU children in their study.

Also [13] who conducted their study on 173 family caregivers and their children with PKU, at the Medical University, Bialystok, Poland, found that, the mean age of PKU children included in the study was 5.5 ± 3.43 years at the time of interview. A statistically significance negative correlation was found between the total practice scores of the family caregivers and the child's age. This is true because the younger the child the more dependent on his family caregivers and this means that the family caregivers need to improve their practices for safe growth & development of these young children.

In the current study, it was found that, approximately both males and females PKU children were equally distributed in this study. The same was reported by several studies [13]; [12] who conducted his study on 186 family caregivers and their children with PKU, at metabolic care center in in Gaza Strip, at Al-Remal PHC in Gaza city and [29] who conducted their multicenter, prospective, non-interventional, observational study conducted in France, Germany, Italy, The Netherlands, Spain, Turkey and the UK on 253 family caregivers and their children with PKU.

[15] conducted a study on 49 family caregivers of PKU children in the Child and Adolescent Consultation Liaison Psychiatry Clinic of children medical center in Tehran for a routine clinical follow-up, reported that, half of PKU children included in all previous studies were females. This is congruent with [9] who stated that, the PKU occurs equally in both males and females. A statistically positive correlation was found between the total practice scores of the family caregivers and the child's sex. This means that, male children still get more concern from the family than females children and this concern increased in case of illness and this could be related to our Egyptian culture and traditions that still differentiate between boys and girls in rural and urban areas.

Regarding the child's age at the detection time of PKU, almost half of the children were detected at the age of 1 to 5 years. This agrees with [12] who found that, most of the cases were discovered before the age of five. This result reflects

lack of community awareness regarding the early detection of PKU and also because PKU screening test is not obligatory applied yet in Egypt as in many other developing countries and also due to the expensiveness of this test. There were a statistically significance negative correlation between the child's age at detection time and the total knowledge & practice scores, since it is not a common disease, so the younger the child's age when the disease was detected, the greater the caregiver's knowledge and practices.

This result was supported by [14] who reported that, family caregiver's knowledge and practices level declines with increasing the child's age at diagnosis. Also, the study performed by [23] on 105 Iranian PKU children and their family caregivers revealed the same results. The current results could means that the younger the children at the detection time the lesser the complications, the mother have the desire and motivation to seek information about the disease. Family caregivers' knowledge is expected to be applied rapidly, so improvement of practices will have occurred, but in contrast, by a late of the detection time, there will be decline in the level of care for the PKU child, as late diagnosis causes more physical & mental complications that could be another barrier for compliance of the caregivers.

Concerning the children's education, it was noticed that, less than one-fifth of the children were enrolled in schools, almost one-third of them were in schools for children with special needs, while the majority of them were not enrolled in any type of schools or did not complete their studies. These findings are incongruent with [16] who conducted a study on 140 family caregivers and their PKU children in the Children Clinic of Metabolic Disorder in Medicine Center in Cairo and found that, less than one quarter of the children were enrolled in schools, most of them were in schools for children with special needs, while the majority of them were not enrolled in any type of schools. On the other hand [30] who conducted a study on 42 cases of classic PKU children in Peking University First Hospital in China and found that a minority of the PKU children were enrolled in schools and their school reports were excellent for those treated since neonates, while the majority of the children either postponed their school enrollment or enrolled in schools for mentally retarded children. Also, these findings are in disagreement with [12] who in a study on children suffering from PKU aged 6-18 years in the Gaza Strip found that, 85.9% of study participants were in prep/primary schools, while 14.1% were in secondary schools, and 15.2% were excellent in school performance, 30.4% were very good, 31.5% were good and 22.8% were weak. This could be related to the most cases of PKU children were discovered recently because PKU screening test is not applied yet in Egypt, in addition to the poor knowledge and practice of the family caregivers of the PKU, therefore, there were many complications, mainly mental and physical disabilities which hindered the child going to school.

The current study indicated that nearly three-quarters of the PKU children had 1 to 3 brothers and sisters. There was a negative correlation between the total knowledge scores of the family caregivers and the child's number of sisters & brothers, which means that the less number of siblings the greater the caregiver's knowledge about the disease. This result is supported by [12] who reported that, the average number of children per family was 2.6. [22] Reported that, the total family caregiver's knowledge scores were higher in families with two or fewer children and lower in large families. Accordingly, the care of the PKU children decreased in large families. It is true that the family caregivers with a large number of children will not have the time and energy for improving their knowledge and practice regarding the disease and its management about the PKU child.

CONCLUSIONS AND RECOMMENDATIONS

The present study concluded that, nearly two-thirds of the family caregivers had poor level of knowledge regarding care of their PKU children, almost quarter of the family caregivers had fair level of knowledge, while minority of the family caregivers had a good level of knowledge, and also there was a considerable unsatisfactory level of practices for the majority of family caregivers regarding care of their PKU children, while only minority of them had a good level of practice

Recommendations: Based on the results of the present study, it can be recommended that: Publication and dissemination of the nursing care protocol in all maternal and child health care centers for families having children with PKU to raise their awareness about the disease management to prevent its complications. Further, research study to be done to investigate the other social and psychological health needs of family caregivers that are evidence-based in order to prevent health problems of children with PKU

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