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Family Caregiver's Knowledge and Practices of Children with Phenylketonuria At Abo El Reesh Hospital

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Abstract

Background: 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.

Aim of this Study : To assess family caregivers' knowledge and self-reported practice regarding Phenylketonuria at the Child clinic of metabolic disorder of the Social Preventive Medicine Center, Abo El-Reesh, Cairo governorate.

Design: A descriptive research design was utilized to fit the purpose of the study.

Setting: The study was conducted at the clinic of metabolic disorder of the Social Preventive Medicine Center, Abo El-Reesh Hospital, Cairo Governorate.

Sample: A sample of 140 family caregivers and their children with Phenylketonuria were included in the study, and the data were collected in six months from October 2011 till March 2012.

Tools for Data Collection: Data was collected using structured interviewing questionnaire, knowledge sheet, and practice sheet (self reported questions) for the family caregiver's home practices regarding PKU.

Results: The study indicates that nearly one quarter of the family caregivers have unsatisfactory total knowledge scores, more than half of the family caregivers have satisfactory total knowledge scores, while minority of the family caregivers have good total knowledge scores. The majority of the family caregivers had a poor level of practice, while only (4.3%) of them had got good level of the practice score, while (3.6%) of the caregivers had fair level. A highly statistically significance 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, as when family caregivers have a good knowledge level regarding their children conditions; this will improve their practices regarding care of their children with PKU.

Conclusion: The study concluded that, the family caregivers had poor level of knowledge and yet the vast majority of children still achieved poor dietary control and poor care.

Recommendations: Based on the study results it is recommended to develop a training program regarding Phenylketonuria to provide them family caregivers with knowledge and skills to help them in caring for their PKU children at home.

Key Words: *Family caregivers – Children – Phenylketonuria.*

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Introduction

CHILDREN are the future of the country, what happens to children in their first days, months and years of life affects their development and also the development of the society. Intervention during the early years can assist in the healthy development of children cognitively, socially, emotionally and physically **[1]**. 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 **[2]**. 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 **[3]**.

Infants with untreated PKU seem to be normal for many months. However, without treatment, phenylalanine (phe) accumulates resulting in mental deficiency, microcephaly, seizures, hyperactivity and purposeless movements, and eczema. In older untreated children the skin and hair are usually fair, the eyes blue and there may be a mousy odor of the skin or urine. 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 function, psychological and behavioral issues, social difficulties, impaired growth, irrita-

bility, and eczema. However, with early and continuing dietary management, children with PKU have normal physical and cognitive development [4].

Over several decades, PKU research has continued to uncover the importance of assessing the family caregiver's knowledge and skills regarding dietary management and adherence at home. Researchers shifted from describing PKU, to focusing on the treatment and prevention of mental retardation associated with untreated PKU which are the responsibilities of the children and their family caregivers [5]. Family caregivers play a pivotal role in the management of childhood Phenylketonuria, this chronic disorder which needs family caregivers' daily effort to deal with the child's disabilities and to maintain a special rehabilitation and diet program. A central issue for parents or other family caregivers is how they deal with the diagnosis, which occurs abruptly soon after the birth and represents a serious threat to the child's development. Moreover, the child's diet is highly restrictive and exacting. Thus, PKU tends to have an intrusive impact on family life, and family caregivers tend to worry about their children's development [6].

Treatment by dietary restriction involves considerable modification of normal eating behavior by the child and cooking behavior by the family caregiver. Motivation to comply with diet depends on psychological factors such as knowledge of the condition, understanding of the treatment goal, reassurance that the diet is working successfully and resistance to peer and other pressures to deviate from the strict regimen. Successful management of PKU is dependent on children and family caregivers. However, children commonly have a difficult time "complying with" or "adhering to" the PKU diet, especially as they move into adolescence given the potentially serious consequences of elevated blood Phe levels, so, it is critical to identify the obstacles to better management faced by children and their family caregivers [7].

Significant of the study :

In Egypt, the study done in Menoufiya Governorate revealed that, 1/3000 (0.03%) which estimates that about 333 neonates are affected with PKU every year as one million babies are born yearly [8]. Furthermore, In Egypt there is few numbers of PKU clinics, only two clinics for all governorates. From the clinical experience the investigator observed that, usually no formal documented system or structure for defining family caregiver's education needs, updating knowledge, or for monitoring education programs and their effects. Poor dietary control is often associated with increasing noncompliance by older children, but it could also be due to a more relaxed dietary approach by the caregivers and increasing dietary errors. Family caregivers understanding of diet therapy and its application has been rarely studied [9].

Consequently, it is possible that deteriorating dietary control in children with PKU may be associated with poor caregiver's knowledge of diet, decreasing motivation, inability to cope with the diet or a combination of all three factors [10].

The family caregivers operate as extensions of health care systems performing complex medical and therapeutic tasks and ensuring care recipient adherence to therapeutic regimens [11]. They operate as home-based "care coordinators" and personal advocates for care recipients. As health care costs and utilization continue to rise, individuals facing physical, mental or behavioral challenges are increasingly dependent on the ability of family or other informal caregivers to operate competently as formal health care providers. Yet, despite their important function in the society, caregivers do not receive adequate training, preparation, or ongoing support from health care systems [12].

The community health nurse has a key role for assessing, planning, organizing and implementing strategies for maintaining high level of knowledge and practice that support the family caregiver of the PKU children [13]. The aim of study was to assess family caregivers' knowledge and practice regarding their PKU children. Results of this study can be utilized to plan and implement educational interventions for improving family caregivers' knowledge and practice regarding their PKU children.

Research questions:

What are the family caregiver's knowledge and practices regarding to PKU?

Material and Methods

Research design:

A descriptive research design was utilized to fit the purpose of the study.

Setting:

The study was conducted at the Child Clinic of PKU metabolic disorder which is affiliated to social preventive medicine center Abo El-Reesh.

Subjects:

All children with PKU attending the clinic and their family caregivers within six months were included in the study, so convenient sample was utilized in this study. Data was collected from October 2011 till March 2012. The total sample after six months was 140 family caregivers.

Tools for data collection:

Three tools were used in this study:

1- Structured interviewing sheet was used to collect data about:

a- Socio demographic characteristics of the PKU child as age, sex, height, weight, education, and ranking between their siblings, etc...

b- Socio demographic characteristics of the caregiver as age, sex, relative degree, education, job, marital status, health status, and family income, etc...

2- Knowledge sheet: Included questions about the caregiver's knowledge regarding PKU as definition, causes, signs and symptoms, treatment and complications, etc...

3- Practice sheet (self reported questions) about the family caregiver's home practices regarding PKU, which included follow-up of the PKU child, monitoring Phe level, the formula preparations, dietary record, diet restriction, etc...

Scoring system:

Knowledge scores were classified as follows:

Complete correct answer was given three points, incomplete correct answer was given two points, and the incorrect answer was given one point. The total scoring points of the knowledge was (69 points) classified as:

- Unsatisfactory level of knowledge=<41 of total score.
- Satisfactory level of knowledge=41-50 of total score.
- Excellent knowledge=51-69 of total knowledge.

Practice scores were classified as follows:

Complete correct answer was given three points, incomplete correct answer was given two points, and the incorrect answer was given one point. The total points of the practice scores was (111 points) considered as:

- Poor level of practice=<66 of total score.
- Fair level of practice=66-83 of total score.
- Good level of practice=84-111 of total knowledge.

Ethical consideration:

- Human subject approval was obtained from the faculty of nursing, ethical committe, Cairo University.
- An official permission was taken from the director of the Social Preventive Medicine Center at Abo El-Reesh.
- Written consent was taken from caregivers who accept to be included in the study.
- The investigator informed each caregiver about the purpose and nature of the study, emphasis that participation in the study is entirely voluntary; anonymity and confidentiality will be assured through coding the data. Every caregiver had the right and freedom not to complete the study process.

Procedures:

Once an official permission was obtained from the director of the Social Preventive Medicine Center at Abo El-Reesh to carry out this study, permission was obtained from the director of clinics, and then permission from the director of metabolic disorder clinic, also acceptance from nursing supervisor was obtained. After explanation of the aim of the study, written and oral informed consent was obtained from all the caregivers who participated in the study, written informed consent was obtained from the representative of the caregiver in absence of reading and writing.

Before distributing the sheets, the investigator informed each caregiver about the purpose and nature of the study, emphasizing that participation in the study is entirely voluntary; anonymity and confidentiality was assured through coding the data. Every caregiver was told that they have the right and freedom not to complete the study process. A pilot study was carried out on 10% of the sample to assess the feasibility of the study as well as clarity and objectivity of the tools, to estimate the average time needed for data collection, to add or omit questions, and to identify varies problems that might be encountered during implementation of the study. No modifications for the questions were done; caregivers who participated in the pilot study were included in the study sample. Data was collected in 6 months from October 2011 till March 2012.

Statistical analysis:

Data were coded, scored, tabulated, and analyzed by compute using the "statistical package for the social science" (SPSS windows) version

11.5. Numerical data were expressed as mean±SD, and range. Qualitative data were expressed as frequency and percentage. Relations between different numerical variables were tested using Pearson correlation. Probability (p-value) less than 0.05 was considered significant and less than 0.001 was considered as highly significant.

Results

The sociodemographic characteristics of the PKU children showed that, the PKU children's mean age was (7.5±4.3) years; more than half of PKU children (52.9%) were males while (47.1%) were females. Concerning the children's education; only (23.6%) of the PKU children were enrolled in schools, while the majority (76.4%) didn't enroll. Regarding the child's age at detection of the PKU, the PKU children's mean age when the disease detected was (4.1 ±3) years (Table 1).

Table (1): Percentage distribution of PKU children according to their age, sex, education and age at detection of the PKU (N=140).

Study variables	Total sample (N=140)	
	Number	%
Age:		
<One year	5	3.6
1-	45	32.1
5-10 years	58	41.4
10 years	32	22.9
X±SD	7.5±4.3	
Sex:		
Male	74	52.9
Female	66	47.1
Education:		
Kindergarten	10	7.1
1st grade primary	1	0.7
2nd grade primary	7	5.0
Special needs schools	13	9.3
1st grade preparatory	1	0.7
2nd grade preparatory	1	0.7
Did not enroll	107	76.4
Age at detection:		
<1 year	17	12.1
From 1 to 5 years	88	62.9
From 6 to 10 years	26	18.6
>10 years	9	6.4
X±SD	4.1±3	

Table (2) showed that, the family caregiver's mean age was (35.8±7.2) years; the majorities (98.6%) of caregivers were females. As regard to the caregivers education, one quarter (25.0%) of the caregivers were not read and write, (30.0%) of them can read and write, (40.7%) of them were having secondary education, while only (4.3%) of caregivers were having university education. Regarding marital status, nearly three quarters (72.9%) of caregivers were married, while (17.1%) were widow and only (10.0%) were divorced. Concerning caregiver's relation to the child, the majority of the caregivers (85.7%) were the mothers of the children, (12.9%) of them were the relatives (sister, grandmother or other relatives, while just (1.4%) of them were the fathers of the children.

Table (2): Percentage distribution of the family caregivers according to age, sex, education, marital status & their relations to the child (N=140).

Sociodemographic characteristics	Total sample (N=140)	
	Number	%
Age/year:		
20–	45	32.1
31-40	62	44.3
>40	33	23.6
X±SD	35.8±7.2	
Sex:		
Male	2	1.4
Female	138	98.6
Education:		
Illiterate	35	25.0
Read & write	42	30.0
Secondary education	57	40.7
University education	6	4.3
Marital status:		
Married	102	72.9
Widow	24	17.1
Divorced	14	10.0
Relation to the child:		
Father	2	1.4
Mother	120	85.7
Sister	6	4.3
Grandmother	5	3.6
Other relatives	7	5.0
Total	140	100.0

More than one third (35.8%) of the caregivers in the urban areas had a good level of total knowledge scores, while only (9.6%) of the caregivers in the rural areas had a good level of the total knowledge scores. As for the satisfactory level of the total knowledge scores, more than half of the caregivers in the rural and urban areas got satisfactory level of the total knowledge scores (56.2%) and (53.7%) respectively. As for the unsatisfactory level of the total knowledge scores, more than one third (34.2%) of the caregivers in the rural areas got unsatisfactory level of the total knowledge scores, while (10.4%) of the caregivers in the urban areas got unsatisfactory level of the total knowledge scores (Fig. 1).

All the caregivers did not have the good level of the total practice scores (0.00%) in the rural areas, while only (8.9%) of the caregivers had a good level of the total practice scores in the urban areas. As for the fair level of the total practice scores, (6.8%) of the caregivers got fair in rural areas, while the caregivers did not have fair level of the total practice scores (0.00%) in the urban areas. As for the poor level, the majority (93.2%) of the caregivers in the rural areas got poor and (91%) of the caregivers in the urban areas got poor (Fig. 2).

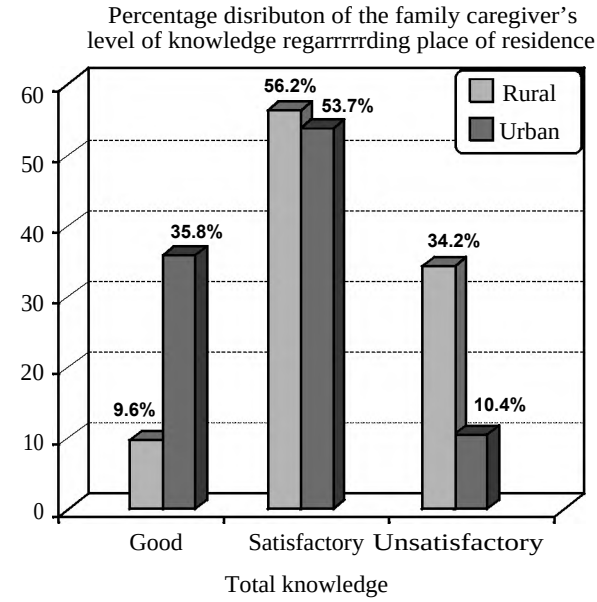

Fig. (1): Percentage distribution of the family caregiver's level of knowledge regarding place of residence (n=140).

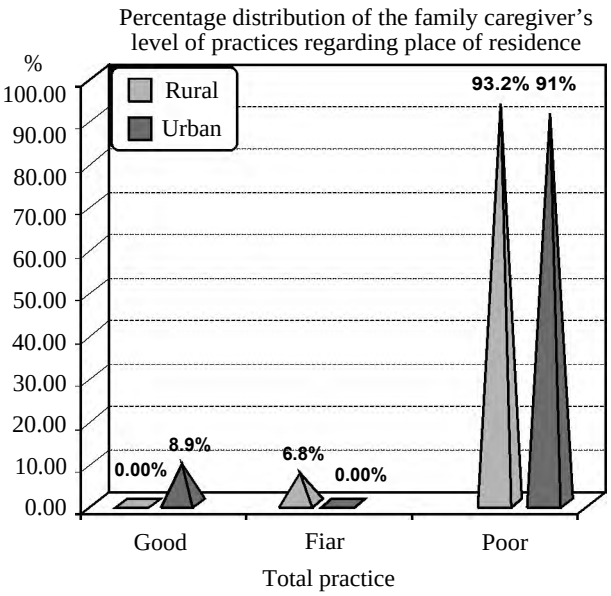

Fig. (2): Percentage distribution of the family caregiver's practices regarding place of residence (n=140).

Table (3) showed that, a statistically significance negative correlation was found between the total knowledge scores of the caregivers and the child's age at detection of the disease (r=0.187*, p=0.027); negative correlation between the total knowledge scores of the caregivers and the child's number of sisters & brothers (r=-0.209*, p=0.021). Regarding the family caregiver's practices, statistically significance negative correlation between the total practices of the caregivers and the child's age (r=0.262**, p=0.002) and highly statistically significance negative correlation between the total practices of the caregivers and the child's age at detection of the disease (r=–0.456**, p=0.000).

Statistically significance negative correlation between the total practice scores of the caregivers and their ages (r=–0.342 * *, p=0.000). As regards to the family caregiver's education, statistically significant positive correlation was found between educational status of the caregivers and total knowledge scores (r=0.457**, p=0.000) and total practice scores as well (r=0.407 * *, p=0.000). Also highly statistically significant negative correlation between the total practices scores of the family caregivers and their marital status (r=-0.221 * *, p=0.009).

A highly statistically significant positive correlation was found between monthly income of the caregivers and their total knowledge scores (r=0.282**, p=0.001). Regarding the place of residence, there were highly statistically significant positive correlation between place of residence of the caregivers and their total knowledge scores (r=0.400* *, p=0.000) and total practice scores as well (r=0.387**, p=0.000).

A highly statistically significance correlation between the caregivers' total knowledge scores and their total practices scores (r=.0.315**, p=0.000).

Table (3): Correlation between the total family caregivers' knowledge and practices scores & the child personal characteristics (N=140).

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 (4): Correlation between the total family caregivers' knowledge and practices scores & their socio-demographic characteristics (N=140).

Caregivers socio-demographic characteristics	Total knowledge		Total practice	
	r	*p*	*r*	*p*
Age	–0.074	0.386	–0.342**	0.000
Education	0.457**	0.000	0.407**	0.000
Marital status	–0.078	0.362	–0.221**	0.009
PKU children in family	–0.123	0.156	–0.206*	0.017
Monthly income	0.282**	0.001	0.054	0.540
Place of residence	0.400**	0.000	0.387**	0.000

** Correlation is highly significant at the level of ≤0.01.
* Correlation is significant at the level of ≤0.05.

Discussion

PKU disease is a common chronic genetic disorder which needs caregiver's daily effort to maintain a special diet program, rehabilitation and to deal with the child's disabilities. Maintaining long-time nutrition regimens and applying complementary diets are the main therapeutic tasks in treating children with PKU which require perfect collaboration of the caregivers **[7]**. Also there is a clear need for more shared education and communication about PKU with respect to all elements of management based on broad consensus guidelines. Increased interchange of knowledge would facilitate better treatment for children within the resources available **[14]**. The purpose of this study was to Assess family caregiver's knowledge and practice regarding Phenylketonuria. One hundred and forty family caregivers and their PKU children were included and three tools were used to collect the pertinent data.

In the present study, it was found that, the majority of the family caregivers were females and were the mothers of the PKU children; these results are supported by a lot of studies **[15,16,17]** where most of family caregivers were the mothers of the children. Hockenberry et al., (2005) **[18]** stated that, mothers of children are considered the most primary health care providers around the world, they takes on enormous responsibilities in providing care and managing of their child's conditions outside the health care institutions. In Egyptian culture, mothers are the main caregivers of their families with or without sick persons at their families because men are busy working.

In the current study, it was found that the mean caregiver's age was 35.8±7.2 years. This goes in the same line with Lord et al. **[6]**, they reported that the mean age of caregivers was 35.3 ±5.2 years, with the same context Gharaei et al. **[17]** reported that the mean age of caregivers was 35.63 ±8.82 years. A highly statistically significance negative correlation was found 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, conversely. This is true because young family caregivers have the strength and the physical ability to care for the children.

Regarding to the family caregiver's education, it was noticed that, more than half of family caregivers were either illiterate or just can read and write and the minority of them had higher education. This finding was supported by Ahmed **[2]**, who found that, almost half of family caregivers were illiterate and the minority of them had higher education. On the other hand, Gallo et al. **[16]** reported that most of the family caregivers completed high school, and approximately one third completed college or graduate school, as well as Lord et al. **[6]**, who found that half of the family caregivers, had completed tertiary education. This difference denotes that most of the females in Egypt are less educated than males as reported by Arab Republic of Egypt **[19]**. 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 **[10,20,21,22]** who reported that, family caregiver's knowledge scores increased with the family caregiver's education levels which may improve the care of the PKU children and family caregiver's illiteracy was a negative factor on dietary compliance. This is true because educated family caregivers usually have more information and better practices compared with illiterate or lower educated family caregivers. As expected, a better adherence was observed with a high level of education of the family caregivers, which could suggest the possibility of specific intervention directed towards less educated families but illiteracy of the caregivers decreased the rate of compliance with the rigid and long-term dietary treatment.

Regarding the marital status, nearly three quarters of family caregivers were married, while the minority were either widow or divorced, which is consistent with many studies **[15,16,23]** 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 practices scores of the family caregivers and their marital status. In spite of slight number of both divorced (14 of 140) and

widow caregivers (24 of 140) in this study, dietary adherence in these cases was significantly poorer than married ones, which is similar to Olsson & Montgomery **[24]**, who found divorced family caregivers had more difficulty in controlling their PKU children's care. This result denotes that child neglect happens more common in divorced families may be because divorced mothers are very busy working and bringing money to support their children.

Concerning the child's age, this study revealed that, the mean age of PKU children included in this study was 7.5±4.3 years. This result is congruent with Lord et al. **[6]**, who reported that, the mean age of the children was 6.6±3.2 years. Also Ozel et al. **[10]**, found that, the mean age of PKU children included in the study was 5.5 ±3.43 years at the time of interview, and also Bilginsoy et al. **[7]**, reported 8.1 ±5.2 was the mean age of PKU children. Regarding the child's age when detecting PKU, almost two thirds of the children aged from 1 to 5 years when the disease was detected. This was agreed with Read, **[23]**, who found that, most of the cases were discovered before 5 years. These results reflected lacking of community awareness regarding early detection of PKU and also because the PKU screening test is not applied yet in Egypt. There was statistically significance negative correlation between the total knowledge scores of the caregivers and the child's age at detection of the disease, which means that the smaller the child's age when the disease is detected the greater the caregiver's knowledge about the disease.

A statistically significance negative correlation was found between the total practices scores of the family caregivers and the child's age, and highly statistically significance negative correlation between the total practices of the family caregivers and the child's age at detection of the disease. This results was supported by Alaei et al. **[22]** and MacDonald et al. **[20]**, who found that, family caregiver's practices level declines with increasing age of the child at diagnosis. A negative correlation was found between age at diagnosis and family caregiver's practice level. It looks that, by a late diagnosis there is a decline in the level of care for the PKU child, as late diagnosis causes more complications including physical and mental retardation that could be another barrier for dietary compliance so younger children had a better dietary control.

More than one third of the family caregivers in the urban areas had a good level of the total knowledge score compared with the minority of them in the rural areas.

More than half of the family caregivers in the rural and urban areas got satisfactory level of the total knowledge scores, while more than one third of the family caregivers in the rural areas got unsatisfactory level of the total knowledge scores compared with the minority of them in the urban areas. This contradicted with Ozel et al. **[10]**, who found that, the total knowledge scores were lower in family caregivers most of them living in cities. Lower knowledge scores among rural caregivers suggested that they did not have knowledge regarding the diet and this knowledge deficit may be due to their lower educational level plus improper health education about their children conditions from the health care team and the limited role of health professionals (including the nurse) in providing the caregivers with the appropriate care of PKU children in rural health facilities.

The current study showed that, there was a highly statistically significance correlation between the family caregivers' total knowledge scores and their total practices scores. This explanation goes in the same line with Ali **[25]**, who mentioned that, when caregivers are provided with the basic knowledge about their children condition, developmental prognosis and various treatment approaches, this will assist them in practicing new and healthy behaviors or also can change unhealthy behavior. Also this opinion is congruent with Macdonald et al. **[20]**, who believed that dietary knowledge is an essential factor on dietary compliance. In consistent with the study results Ozel et al. **[10]**, who reported that, the caregiver's practice scores and dietary compliance was much lower when knowledge scores was particularly low. These results mean that, the family caregivers' knowledge had an effect on their practices, as when family caregivers have a satisfactory knowledge level regarding their children conditions; this will improve their practices regarding care of their children with PKU.

There were negative correlation between the total knowledge scores of the family caregivers and the child's number of sisters & brothers, which means the less number of sons' the greater the caregivers knowledge about the disease. This result is supported by Ozel et al. **[10]** and Alaei et al. **[22]**, who reported that, 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 diminished in families with many people. This is true because when family caregivers had a lot of children at their families they will not have enough time and energy to read and know about the disease and its management.

The current study revealed that, there was statistically significance negative correlation between the total practice scores of the family caregivers and number of PKU children in the family, this is similar to Alaei et al. **[22]**, who reported that, by increasing number of affected children in the family they had poor Phe diet control and increase in blood phenylalanine concentrations. It seems that, by increasing the affected children in the family there will be more pressure on the family resources physical, psychological and/or financial, because in Egypt special foods or dietary supplementations are not available for free.

Conclusion:

This study was undertaken to assess family caregiver's knowledge and practice regarding their PKU children at the Social Preventive Medicine Center at Abo El-Reesh Hospital. The present study concluded that, nearly one quarter of the family caregivers had unsatisfactory total knowledge scores regarding their PKU children, more than half of the family caregivers had satisfactory total knowledge scores, while minority of the family caregivers had a good total knowledge scores, and also there was a considerable poor level of practices for the majority of family caregivers regarding their PKU children, while only (4.3%) of them had a good level of the score practice, and (3.6%) of the family caregivers had fair level score.

Recommendations:

In the light of the findings of this study, the following recommendations are suggested for improving the care offered by the family caregivers to their PKU children.

1- Health education Programs for raising the awareness of the family caregivers regarding the available referral center providing care and follow-up for PKU children through different types of mass media such as, radio, television or internet.

2- Further researches are needed to explore more about the nursing strategies for caring of the child with PKU.

References

1- DONALD L. WONG, MARILYN J. HOCKENBERRY and DAVID W.: Nursing care of infant and children, 8th ed., Mosby Comp., Pp. 900, 901, 2006.

2- AHMED S.: Home care offered by family caregivers to preschool children, suffering from hemiplegic palsy. Minia University pp. 81, 2009.

3- Robert Wood Johnson Foundation: Commission to Build a Healthier America: America's Health Starts With Healthy Children: How Do States Compare? Available at: www.commissiononhealth.org/statedata, 2008.

4- GASSIO R., ARTUCH R. and VILASECA M.A.: Cognitive functions in classic Phenylketonuria and mild hyperphenylalaninemia: Experience in a pediatric population. Dev. Med. Child. Neurol., 47: 443-448, 2005.

5- BOSCH A.M., TYBOUT W. and VAN SPRONSEN F.J.: The course of life and quality of life of early and continuously treated Dutch patients with Phenylketonuria. Journal Inherit Metab. Dis., 30 (1): 29-34, 2007.

6- LORD B., UNGERER J., and WASTELL C.: Implications of Resolving the Diagnosis of PKU for Parents and Children, Journal of Pediatric Psychology, Vol. 33, No. 8, pp. 855-866, 2008 Published by Oxford University Press on behalf of the Society of Pediatric Psychology. Journals.Permissions@oxfordjournals.org.

7- BILGINSOY C., WAITZMAN N., LEONARD C.O. and ERNST S.L.: Living with Phenylketonuria: Perspectives of patients and their families, J. Inherit Metab. Dis., 28: 639-649, 2006.

8- EL ARABY H., FATEEN E. and GOUDA A.: Screening for Phenylketonuria and galactosemia among Egyptian newborns in Menoufiya governorate Egypt. J. Med. Hum. Genet., Vol. 10, No. 2, Nov., 2009.

9- BEKHOF J., VAN SPRONSEN F.J., CRONE M.R., VAN RIJN M., OUDSHOORN C.G. and VERKERK P.H.: Influence of knowledge of the disease on metabolic control in Phenylketonuria. Eur. J. Pediatric., 162: 440-442, 2003.

10- OZEL H.G., KUCUKKASAP T., KOKSAL G., KALKANOGLU SIVRI H.S., DURSUN A., TOKATLI A. and COSKUN T.: Does maternal knowledge impact blood phenylalanine concentration in Turkish children with Phenylketonuria? References to electronic databases: PKU: OMIM 261600. HPA: OMIM 261600. SSIEM and Springer, 2008.

11- MASLOW K., LEVINE C. and REINHARD S.: Assessment of family caregivers: A public policy perspective. In Voices and Views from the Field (Vol. II). Report from a National Consensus Development Conference. San. Franci., 2006.

12- SHEPHERD J. and HOLLANDER M.: Building Awareness of the Safe Use of Medications among Family Caregivers of Children with Chronic Illness: Synthesis of the Literature (English) ISBN 1-896870-33-3, 2006.

13- LUNDY K.S. and JANES S.: Community Health Nursing, Caring for the Public's Health (2nd ed., pp 94, 95), Jones & Bartlett Pubs, 2009.

14- MICHALS-MATALON K.: Sapropterin dihydrochloride, 6-R-L-erythro-5, 6, 7, 8-tetrahydrobiopterin, in the treatment of Phenylketonuria. Expert Opin Investig Drugs, 17: 245-51, 2008.

15- IEVERS-LANDIS C.E., HOFF A.L., BREZ C. and CANCILLIERE M.K.: Situational Analysis of Dietary Challenges of the Treatment Regimen for Children and Adolescents with Phenylketonuria and Their Primary Caregivers. Vol. 26, No. 3, June, Copyright # by Lippincott Williams & Wilkins, Inc. Printed in U.S.A., 2005.

16- GALLO A.M., KNAFL K.A. and ANGST D.B.: Family information management patterns in childhood genetic conditions. Journal of Pediatric Nursing, Vol. 24, No. 3 (June), Elsevier Inc., 2009.

17- GHARAEI J.M., MOSTAFAVI S. and ALIREZAEI N.: Quality of Life and the Associated Psychological Factors in Caregivers of Children with PKU, Iran J. Psychiatry. Spring, 6 (2): 66-69. PMCID: PMC3395945, 2011.

18- HOCKENBERRY M., WILSON D. and WINKELSTEIN M.: Wong's Essential of Pediatric Nursing. 7th ed., Mosps, Elsevier, India, pp. 550-555, 2005.

19- Arab Republic of Egypt On Literacy and Adult Education, online at http://www.unesco.org/ fileadmin/ MULTIMEDIA/INSTITUTES/UIL/ confintea/ pdf/ National_Reports/ Arab%20States/Egypt.pdf, 2008.

20- MACDONALD A., DAVIES P., DALY A., HOPKINS V., HALL S. K., ASPLIN D., HENDRIKSZ C. and CHAKRAPANI A.: Does maternal knowledge and parent education affect blood phenylalanine control in Phenylketonuria? Journal of Human Nutrition and Dietetics, 21, pp. 351-358, 2008.

21- COTUGNO G., NICOLÒ1 R., CAPPELLETTI S., GOFFREDO BM., DIONISI VICI C. and CIOMMO V.: Adherence to diet and quality of life in patients with Phenylketonuria. The Author(s)/Acta Pediatric Foundation Acta. Pediatric., 100, pp. 1144-1149, 2011.

22- ALAEI M., TOTONCHI G. A., GACHKAR L. and FARIVAR S.H.: Family Social Status and Dietary Adherence of Patients with Phenylketonuria Iranian Journal of Pediatrics, Volume 21 (Number 3), September, Pages: 379-384, by Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences, 2011.

23- READ C.Y.: Using the impact of event scale to evaluate psychological response to being a Phenylketonuria gene carrier. J. Genet. Couns., 13: 207-219, 2004.

24- OLSSON G.M., MONTGOMERY S.M. and ALM J.: Family conditions and dietary control in Phenylketonuria. Journal Inherit Metab. Dis., 30: 708-715, 2007.

25- ALI N.: Needs of the physically handicapped adults in the vocational rehabilitation centers, Master thesis, Faculty of Nursing, Alexandria University, pp. 25, 2008.