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Knowledge, Attitudes and Reported Practices for Mothers of Phenylketonuria Children at Hereditary Guideline Center in Assiut Governorate

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Abstract
Background: Phenylketonuria is an inherited autosomal recessive metabolic disorder that causes elevated blood phenylalanine concentrations. **Aim of the study:** To assess knowledge, attitude and reported practices of mother with phenylketonuria children at Hereditary Guideline Center in Assiut Governorate. **Methods:** Descriptive research design was used in this study. The total sample number of the studied sample was 360 mothers. Convenience sample was used **Tools of the study:** personal interview questionnaire which included Part (1) socio-demographic, Part (2) questions to determine knowledge, Part (3) attitude and Part(4) reported practices for mothers of phenylketonuria children. was used **Results:** slightly more two fifths (42.8%) of mothers aged 30-<40 years, 31.7% of them had secondary education and 52.5% of them had low social class. According to their knowledge, 18.3% of the studied mothers had good knowledge regarding phenylketonuria, while 40% of them had fair knowledge and 41.7% had poor knowledge. Moreover, 81.4% of mothers had negative attitude toward phenylketonuria, and 76.1% of them had unsatisfactory practice. **Conclusion:** Most of the studied mothers had Poor knowledge, negative attitude and unsatisfactory practices **Recommendations:** Health education programs regarding phenylketonuria should be provided for mothers to improve their knowledge, attitude and practice.

Keywords: Attitude, Hereditary, Knowledge, Phenylketonuria & Practice

Introduction:
Phenylketonuria (PKU) is an inherited metabolic disorder caused by an enzyme deficiency (phenylalanine hydroxylase) (PAH; 612349), resulting in accumulation of phenylalanine and its metabolites in the blood. PKU has autosomal recessive inheritance; consanguineous marriage is an important risk factor for phenylketonuria. Both parents must carry the faulty gene. Mutations of the PAH gene result in decreased activity to complete inactivation of the PAH enzyme lead to accumulation of excess phenylalanine in the blood and can cause severe and irreversible intellectual disability if untreated **(Wesonga et al., 2016; Saudubray and Garcia-Cazorla 2018; Hofman et al., 2018; Said & Draz 2019; Shoraka et al., 2020) .**

The incidence of PKU varies among geographic regions worldwide. In Europe, the highest incidence has been observed in Ireland at a rate of 1: 4,500. It is also common in few parts of China, while it is rarely observed in African nations. In Turkey, an incidence as high as 1: 2,600 has been reported **(Ford et al., 2018).** In Menoufiya governorate prevalence was 1/3000 **(Morad et al., 2019).** while, in Fayoum Governorate, the incidence rate during 2017 was 2.8:10000. identified incidence rate in Egypt as a whole by 1 per 7500 (i.e. 1.3 in 10000**) (Gad et al., 2019).**

The Egyptian Ministry of Health and Population was not conduct A National Screening Program for PKU until November 2015.So,the community had a little knowledge, attitude and practice toward phenylketonuria .More studies prepared for that, in Cairo University the majority of study had poor knowledge and practice level **(Abd-Elkodoos et al., 2018a).** Moreover, **Witalis et al.,2016** had a negative attitude. Also, **El-sayed et al., 2020** at Ain Shams University Hospital represent that more than half of the mothers had poor knowledge and practice.

Children with untreated PKU appear normal at birth but by age 3 to 6 months, they begin to lose interest in their surroundings. 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. By age one year, children are developmentally delayed. It is usually severe and most children require institutional care if the condition remains untreated **(Khaton, 2016).**

Treatment strategies for PKU aim at lowering blood Phe concentration by dietary and/or pharmacologic therapy.

Dietary therapy consists of a Phe restricted diet in combination with Phe-free or Phe- restricted L-amino acid-rich medical foods and/or modified low-protein foods **(Ania et al., 2021).** The newborn Screening program is a public importance health measure aimed at identification of early cases, management of afflicted infants, and making efforts to reduce morbidity and mortality among newborns by screening, follow up, diagnosis, treatment/management, and evaluation **(Twfeeq & Abed, 2016).**

Community health nurse as a primary health care nurse plays an important role in PKU children assessment and provides health education to the population about the disease include: early case finding to prevent complication, nutrition and diet restricted and referral for treatment of Phenylketonuria disease. They provide a premarital counseling about consanguineous marriage to determine if parents carry the gene for PKU during preconception period **(Khaton, 2016).**

Provide additional support and resources and regular contact outside of scheduled visits can maintain engagement with PKU children. Also, Follow-up calls or e-mails after each clinic visit can help assess a children with PKU**(Abd-Elkodoos et al., 2018b).**

Significance of the study:

The national program for the early detection of disability diseases in newborns conduct two pilot studies in both 2012 and 2013 to assess the prevalence of the disease by random samples from 9 governorates in Egypt and the results confirmed high incidence of PKU 1/7000. On other hand there was a preliminary Study in Sohage was reported in this study that include 18,000 cases seen in the pediatric clinics of Sohag University hospital over three years, one hundred were suspected and 2 cases were confirmed to have PKU. This prevalence of 1 in 9000 is higher than what was reported by other groups. However, increases in the number of diagnosed cases over time have been reported. In addition , the majority of PKU children had intellectual disability, ADHD symptoms, and autism **(Sadek et al., 2018 & Morad et al., 2019).**

Untreated children with PKU include behavioral and psychological problems such as; hyperactivity, irritability, inability to sit, sleep disturbances, psychomotor agitation, temper tantrums, uncontrollable attacks of rage, short attention span, aggressive behavior, poor ability to follow directions, poor ability to learn new things, psychotic behavior, severe behavioral disturbance, destructiveness and self-injury, tremors, muscular hyper tonicity, whole body repetitive movements, paraplegia, as well and quadriplegia, developmental delay or low self-esteem, Others complications include, eczema (can involve large areas of the body); sensitivity to sunlight and very light skin (due to excess PHE inhibiting melanin formation) body odor identified as "musty" **(Fouad & Elmoneem,2016; Qu et al.,2019; Elsayed et al., 2020).**

Aim of the study:

To assess knowledge, attitude and reported practices of mother who had phenylketonuria children at center of hereditary guideline in Assuit Governorate.

Research questions:

a. What is the mothers' level of knowledge with PKU children regarding care of Phenylketonuria children in Assuit Governorate?
b. What is the mothers' attitude regarding Phenylketonuria children in Assuit Governorate?
c. What is the mothers' reported practice regarding care of children with PKU in Assuit Governorate?

Research design:

Descriptive research design was applied in this study.

Setting of the study:

The study was carried out at hereditary guideline centre in which cited nears to Dictorate of health in Assuit Governorate. The hereditary guideline centre serve patient with Autism, Down syndrome and dairy allergy. The number of daily work hours in the center 8 hours/shift. The number of the employee was 18 who classified as 4 physicians (pediatrician and nutrional specialist), 5 pharmacologists, 5 nurses, 3 administrators, laboratory technician, and one cleaning worker.

Sample:

Convenience sample was used. The sample was 360 mothers with ages ranged from 18-57 years.

Tools of the study:

After reviewing the related literature, the structured interview questionnaire was developed by the researcher to collect data from mothers with phenylketonuria children which included four parts:

Part (1): Socio demographic characteristic

Personal data was involved 10 open ended questions as age of the child, number of sisters & brothers, child rank, number of years between this child and previous child. PKU child assessment when discovered of disease and current weight, height and head circumference and age when disease detection **(Fouad and Elmoneem 2016; Kucukkasap Comert andKoksal, 2017).** Also, socioeconomic scale was used which covered 16 closed-ended questions. It classified into 7 domains, the first domain assessed the different levels of education and cultural, the 2nd domain involved questions related to different types of occupation, 3rd domain related to family numbers, 4th domains are related to the family possessions, 5th domains covered the economic status and source of income, monthly income, 6th domains assessed the

home sanitation, and finally 7th assessed the health care **(El-Gilany & El-Wasify 2012).** Socio-economic level divided into: very low: <42, low: 63-<42, middle: 71-<63 and \high: 84-<71.4.

Part (2): Knowledge of the studied mother regarding phenylketonuria

It included 16 questions which divided into three sections: the first section involved 6 questions about the mothers' knowledge regarding PKU disease as definition of PKU, risk factor, causes, early symptoms, latent symptoms and complications of phenylketonuria. The second section contained 4 questions about artificial feeding such as temperature that keep and duration for use of free phenylalanine formula, period of changing nipple of feeding bottle and lastly kind of pot to boil water. The last section had 5 questions related to the supported services as source of nutritional plan, prohibited foods, contraindicated ,type of medications and finally one question about the social support **(Abd-Elkodoos et al., 2018a;Cazzorla et al., 2018; Borghi et al., 2020 & Elsayed et al., 2020).**

Scoring system of knowledge:

The total score for the knowledge was equal 37 marks; answers of questions were either correct or incorrect, a score of one was given for correct answer and zero was given for an incorrect answer and I don't know. There were questions with more than one answer. Each item summed up and then converted into a percent. Mothers knowledge was classified into Poor level of knowledge = < 50% of total score, Fair level of knowledge= 50% to 70% of total score and good knowledge =more than 70% of total knowledge **(Abd-Elkodoos et al., 2018a).**

Part (3): The attitude of the studied mothers toward PKU disease

It was developed by the researcher based on the relevant literature to assess the mothers' attitude toward PKU disease. It was contained 14 statements. The response to the statements measured based on 3 points Likert scale (agree, uncertain and disagree). Items were scored (0, 1 and 2) respectively for reversed negative statements. There was six reversed questions which included (Do you think that genetic diseases prevalent in girls more than in boys, Children with genetic diseases do not enjoy a normal healthy life, A child with genetic diseases is disabled child, A child with genetic diseases that you are ashamed of in front of your family, From the point of view of society, this disease is considered a stigma and Do you think that you will have children with the same disease in the future). While the rest of questions were scored (2, 1, 0) respectively **(Witalis et al. 2017& Khdair et al. 2021).**

Scoring system of attitude:

The total score equal 28 marks; each item summed up and then converted into a percent .Mothers attitude was classified into positive attitude (60% and more) or negative attitude (< 60%) for the studied mothers **(Witalis et al. 2017).**

Part (4): Checklist of the reported practice for the Studied mothers:

It included 30 statements in three sections: Formula preparation (4 statements), feeding process (2 statements), dietary record (4 statement), making baked goods for the PKU children (11 statements), follow up (1 statement), hair and nail care (2 statements), hand wash (2 statements), clothing and bathing (2 statements), and skin care (2 statements) **(MacDonald et al., 2016; Abd-Elkodoos, et al., 2018a; Elsayed et al.,2020).**

Scoring system of reported practice:

The total score was 30 marks; mark for each statement. Answers of the checklist were either done or not done, with the score of one mark for done and zero for not done. Each item summed up and then converted into a percent. Mothers' practices were classified into either satisfactory (60% and more) or unsatisfactory (< 60%) **(Fouad and Elmoneem 2016; Abd-Elkodoos et al., 2018a).**

Validity:

Content validity for the questionnaire was examined and revised by 5 experts in nursing science in Assuit University to confirm that questionnaire was cleared and relevant to PKU.

Reliability:

Reliability of the tools was tested using Cronbach's Alpha for knowledge, attitude and practice (0.823, 0.743& 0.869) respectively and El-Gilany Socio-economic scale was 0.66.

Data collection was carried out in three phases:

Preparatory phase:

It included reviewing of the current literature in the various aspects of the review using text books, articles, different studies, internet and journals in order to develop the study questionnaire for data collection.

An official approval letter was obtained from the Dean of the Faculty of Nursing, Assiut University, to the heath Directorate (department of maternities and childhoods) and the director of metabolic disorder clinic to conduct the study.

Pilot study:

Pilot study was carried out before starting data collecting on 10% from the total of sample (36 cases) that included in the study. The time of pilot study collected in the mid of October 2020.The aim of this study was to test the clarity of the tools and estimate the required time to fill the questionnaire based on the

results of pilot study and no modifications in the tools were done.

Data collection phase (Field work):

Data collection started from January 2021 to July, 2021; two days/ week for the studied mothers and approximately from 8-9 questionnaires were collected daily. Filling of the questionnaire was taken from 25-30 minutes.Verbal consent was obtained from all the studied mothers before the study enrollments, after that a detailed explanation for the study objectives was done.

Ethical consideration:

Research proposal was approved from Ethical Committee at the Faculty of Nursing –Assuit University, there is no risk for the studied mothers during the application of the research, the study followed the common ethical principles in clinical research, and oral agreement was taken from them after explaining the nature and the aim of the study and they have the right to refuse or to participate. Study participant's privacy was considered during data collection.

Statistical Design:

- Upon completion of data collection, the data was reviewed, prepared for computer entry, coded, scored, tabulated, and analyzed by computer using the "statistical package. Descriptive statistics (i.e., frequencies, percentage ….etc.) was done using computer program SPSS version 20.
- Frequency and percentage were used for numerical data also, fisher exact test was used.
- Chi-square test used to compare differences in the distribution of frequencies among different groups. It is considered * significant when P-values were less than 0.05 or (P< 0.05).

Result:

Table (1): Socio demographic characteristic of the studied mothers with PKU children at hereditary guideline center in Assuit Governorate, 2021

Socio demographic characteristic	No. (360)	%
Mother age: (years)		
< 30	144	40.0
30 - < 40	154	42.8
≥ 40	62	17.2
Mean ± SD (Range)	32.16 ± 7.70 (18.0-57.0)	
Child age(years):		
< 2 years	106	29.5
2 – 5 years	152	42.2
> 5 years	102	28.3
Educational level:		
Illiterate	89	24.7
Basic education	111	30.8
Secondary education	114	31.7
University degree or equivalent	44	12.2
Post university	2	0.6
Mothers' working condition:		
Housewives	339	94.2
Working	21	5.8
Place of residence:		
Urban Slum	36	10.0
Rural	279	77.5
Urban	45	12.5
Social class level:		
Very low	68	18.9
Low	189	52.5
Moderate	75	20.8
High	28	7.8

Table (2): The current medical history of PKU children at hereditary guideline center in Assuit Governorate, 2021

Items	No. (360)	%
PKU child medical history:		
Yes	96	26.7
No	264	73.3
Autism	5	5.2
Increased brain electricity	7	7.3
Brain atrophy	17	17.7
Epilepsy	3	3.1
Convulsion	22	22.9
Motor delay	72	75.0
Speech problems	12	12.5

#More than one answer

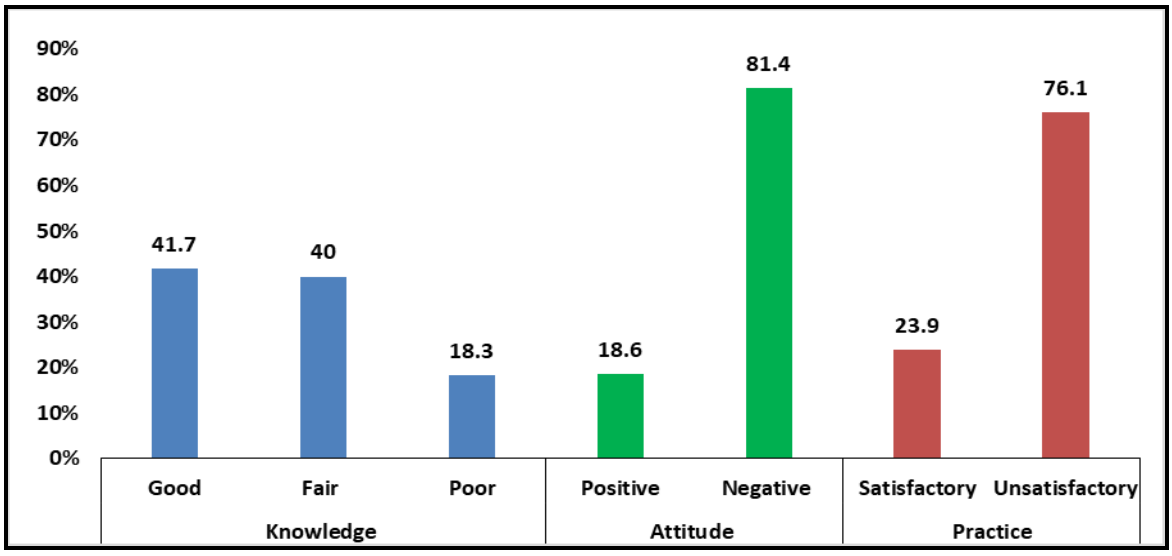

Figure (1): The studied mothers' level of knowledge, attitude and practices regarding PKU at hereditary guideline center in Assuit Governorate

Table (3): Relationship between the mothers' total knowledge about PKU and personal data at hereditary guideline center in Assuit Governorate, 2021

Personal data	Knowledge level						P-value
	Poor (n= 150)		Fair (n= 144)		Good (n= 66)		
	No.	%	No.	%	No.	%	
Age: (years)							
< 30	54	36.0	62	43.1	28	42.4	0.076
30 - < 40	66	44.0	66	45.8	22	33.3	
≥ 40	30	20.0	16	11.1	16	24.2	
Educational level							
Illiterate/ Read & write	67	44.7	30	20.8	4	6.1	
Basic education	42	28.0	32	22.2	25	37.9	0.000*
High school/ Intermediate institute	34	22.7	76	52.8	16	24.2	
Faculty/ postgraduate	7	4.7	6	4.2	21	31.8	
Mothers' job							0.000*
Housewife	149	99.3	138	95.8	52	78.8	
Working	1	0.7	6	4.2	14	21.2	

Personal data	Knowledge level						P-value
	Poor (n= 150)		Fair (n= 144)		Good (n= 66)		
	No.	%	No.	%	No.	%	
Residence							
Popular urban area	17	11.3	12	8.3	7	10.6	0.017*
Countryside	121	80.7	115	79.9	43	65.2	
Attend classy	12	8.0	17	11.8	16	24.2	
Social class:							
Very low	51	34.0	15	10.4	2	3.0	0.000*
Low	72	48.0	87	60.4	30	45.5	
Moderate	24	16.0	34	23.6	17	25.8	
High	3	2.0	8	5.6	17	25.8	

**P Value = (0.000). Chi-square and Fisher exact test.*

Table (4): Relationship between the mothers's total attitude about PKU and personal data at hereditary guideline center in Assuit Governorate, 2020

Personal data	Attitude level				P-value
	Negative(N=293)		Positive(N=67)		
	No.	%	No.	%	
Age: (years)					
< 30	115	39.2	29	43.3	
30 - < 40	132	45.1	22	32.8	0.120
≥ 40	46	15.7	16	23.9	
Educational level:					
Illiterate/ Read & write	97	33.1	4	6.0	
Basic education	74	25.3	25	37.3	0.000*
High school/ Intermediate institute	109	37.2	17	25.4	
Faculty/ postgraduate	13	4.4	21	31.3	
Mother's job:					
Housewife	286	97.6	53	79.1	0.000*
Working	7	2.4	14	20.9	
Residence:					
Popular urban area	29	9.9	7	10.4	
Countryside	235	80.2	44	65.7	0.007*
Attend classy	29	9.9	16	23.9	
Number of family members:					
< 5	120	41.0	33	49.3	0.215
≥ 5	173	59.0	34	50.7	
Social class:					
Very low	66	22.5	2	3.0	
Low	158	53.9	31	46.3	0.000*
Moderate	58	19.8	17	25.4	
High	11	3.8	17	25.4	

**P Value = (0.000).Chi-square test and Fisher exact test.*

Table (5): Relationship between the mothers' total practice about PKU and personal data at hereditary guideline center in Assuit Governorate, 2020

Personal data	Practice level				P-value
	Unsatisfactory(N=274)		Satisfactory(86)		
	No.	%	No.	%	
Age: (years)					
< 30	105	38.3	39	45.3	
30 - < 40	127	46.4	27	31.4	0.037*
≥ 40	42	15.3	20	23.3	
Educational level:					
Illiterate/ Read & write	101	36.9	0	0.0	
Basic education	91	33.2	8	9.3	0.000*
High school/ Intermediate institute	77	28.1	49	57.0	
Faculty/ postgraduate	5	1.8	29	33.7	
Mother's job:					
Housewife	273	99.6	66	76.7	0.000*
Working	1	0.4	20	23.3	
Residence:					
Popular urban area	19	6.9	17	19.8	
Countryside	234	85.4	45	52.3	0.000*
Attend classy	21	7.7	24	27.9	
Number of family members:					
< 5	108	39.4	45	52.3	0.035*
≥ 5	166	60.6	41	47.7	
Social class:					
Very low	68	24.8	0	0.0	
Low	179	65.3	10	11.6	0.000*
Moderate	24	8.8	51	59.3	
High	3	1.1	25	29.1	

**P Value = (0.000).Chi-square test and Fisher exact test.*

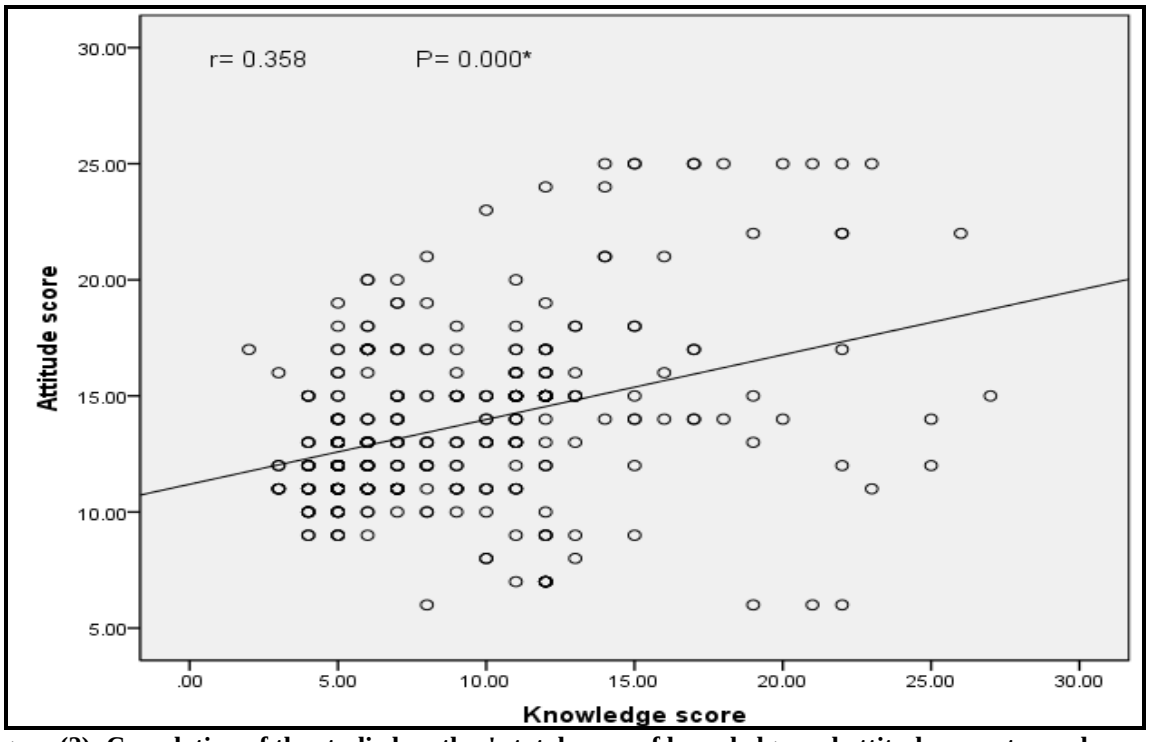

Figure (2): Correlation of the studied mother's total score of knowledge and attitude score toward care of their PKU children

Table (1): Regarding socio demographic characteristic of the studied mothers with PKU children, table (1) showed that 42.8% of mothers aged 30 - < 40 years, 77.5% of them was rural, and 10% were living in urban slum. Regarding their social class, 52.5% of the studied sample was low social class, while 18.9 % very low and 7.8% in high social level. Moreover, it showed 24.7% of mothers were illiterate, 31.7% had high school or equivalent while 12.2 % had university and post graduate.

Table (2): Showed that 26.7% of them had chronic diseases as: 75.0% motor disability, 22.9% convulsion, 17.7%brain atrophy, 12.5% speech problem, 7.3%increased brain electricity, 5.2%, Autism, 3.1% epilepsy.

Figure (1): Showed that 41.7% of the studied mothers had poor knowledge regarding PKU while 40% of them had fair knowledge. Also, 18.3% had good knowledge regarding PKU, Moreover, the figure displayed that 18.6% of the studied mothers had positive attitude toward PKU disease, versus 81.4% had negative attitude. Moreover, 23.9% of the studied mothers had satisfactory level of practice about PKU disease.

Table (3): Showed that the studied mothers had highly significant relationship between knowledge and their educational level, working condition and social class and P value = (0.000). Also, there was a statistically significant differences between mother's knowledge and their residence (P value =0.076).

Table (4): Revealed that there was statistically significant between mothers attitude and their educational level, working condition, and social class (P Value= 0.000). There was statistically difference between mother's attitude and residence (0.007) respectively.

Table (5): The table illustrated that there was statistically significant relationship between mothers practice and their Age, educational level, working condition, residence and social class ((P value=0.000). respectively. There was statistically difference relationship between mothers practice and their Age (P value= 0.037).

Figure (2): Showed that there was correlation between knowledge score and attitude score. It was found that there was a significant positive correlation between Knowledge and practice scores (P value=0.000 r=0.383).

Discussion:

Phenylketonuria disease is a common chronic genetic disorder that needs the caregiver's daily effort to maintain a special diet program, rehabilitation, and dealing with the child's disabilities **(Elsayed et al., 2020)** Phenylketonuria (PKU) still poses a therapeutic challenge for patients and medical professionals **(Witalis et al., 2016).**

The current study revealed that more than one-quarter of the studied children their ages were < 2 years while more than two-fifths their ages ranged from 2-5 years. Also, more than one-quarter of them had >5 years. Regarding the age of mothers with PKU children, two-fifths of the studied mothers were < 30 yrs. while more than two-fifths ranged from 30-< 40 yrs. Also, less than one-fifth of them ≥40 yrs., with the mean range of age 32.16 ± 7.70.

According to the level of mother's education, less than one-quarter of them weren't joined for education, more than one-quarter had secondary educational level. Also, less than one-fifth had university and post-graduate education. The study results were supported by **Abd-Elkodoos et al., 2018a** who mentioned that more than one quarter of them cannot read and write, less than one third can read and write, less than one third had secondary education, while only twelve percent had university education**.** Also, **Fouad & Elmoneem, 2016** stated that more than one-tenth of caregivers were illiterate, and more than one-tenth of them can read and write, almost two-fifths of them had intermediate education, while only less than one-fifth of them were having a university education. This result is opposed to **Witalis et al., 2017** who mentioned that more than two-thirds of parents had vocational or secondary education, one quarter had higher education and small size of the sample had primary school education.

Related to the mothers' working conditions, the majority of them were housewives. This result was supported by **Fouad and Elmoneem,2016** who conducted a study titled "nursing intervention program for family caregivers having children with phenylketonuria" and found that the majority of caregivers weren't working.

According to the place of residence, slightly less than three-quarters of the studied mothers are living in the rural area. These results were supported by **Elsayed et al.,2020** who mentioned that more than one fifth of them lived in rural areas. The study contrast with, **Gharaei et al., 2012.** who stated the difference could be related to the socioeconomic and cultural factors .In my opinion that most of females in Egypt are less educated than males and this is more common among rural than urban females as reported by central agency for public mobilization and statistics, literacy and adult educations.

According to the social level, less than one-fifth of the studied mothers had very low social class and more than one-fifth had moderate level and the minority was high social class level. These findings are in contrast with **Witalis et al., 2017** who conducted their study on family caregivers and their

children with PKU, at the Medical University, Bialystok, Poland, and found that less than three-quarters of them described their family's economic conditions as very good or good, and one fifth as average and poor.

As regarding the medical history of PKU children, the result concluded that less than one fifth had a medical history of brain atrophy, slightly more than one fifth had a convulsion, three-quarters of them had a motor delay, and less than one fifth had a speech problem. The current study is similar to **Abd-Elkodoos, et al., 2018** who mentioned that more than two-fifths had increased brain electricity, less than one-fifth had autism and convulsion. Also, These results were in the same line with **Gad et al., 2019** who stated that the children had a history of motor developmental history, motor delay, convulsion, and autism. Also, **Roby et al., 2021** revealed that the disease affected less than half of children's general health such as delayed growth of weight and length, behavioral problems, delayed social and intellectual skills, skin related disease, convulsions, and bad odor of the body. The current study represent that PKU is apparent to be more prevalent among the children with high consanguinity among parents.

The current study showed that slightly more than two fifths had poor knowledge, while two fifths of them had fair knowledge regarding PKU and less than one-fifth of the studied mothers had good knowledge. This amplification goes in the same line with **Elsayed et al., 2020** Elsayed et al.,2020 Elsayed et al.,2020 who mentioned that more than half of mothers had poor knowledge about PKU. Also, **Abd-Elkodoos et al., 2018** stated that slightly less than two thirds had a poor level of knowledge regarding PKU, more than one-fifth had fair knowledge and less than one-fifth had good knowledge regarding PKU.

Regarding the mothers' attitude, the study detected that the majority of the mothers with PKU children had a negative attitude. This result is supported by **Elsayed et al., 2020** who showed the majority of mothers had a negative attitude to phenylketonuria. On the other hand, the results contrast with **Witalis et al., 2016** who demonstrated that more than three-fifths of parents did not accept the disease. The researcher in this study that negative attitude with Phenylketonuria related to that this program not applied even 2015.

The present study showed that there was a statistically significant difference between mothers' total practice and their occupation, this result differed with **Abd-Elkodoos et al., 2018** who revealed that no statistically significant correlation was found between family caregiver's job and their total practice & knowledge scores.

The mother's knowledge had an effect on meet needs and practices toward their children, if mothers have a good level of knowledge toward their children's cases; this will improve their practices to PKU children **(Abd-Elkodoos et al. 2018b & Elsayed et al., 2020).**

The study revealed that there was a highly statistically significant between educational status especially illiterate and basic education of the mothers and total knowledge, attitude, and practice scores, which was supported by **Witalis et al., 2017 & Abd-Elkodoos, et al., 2018a** who reported that family caregiver's knowledge scores increased with the family caregiver's educational levels. In addition to this, the result difference with **Alaei et al. 2011** who found that no relation between the family caregiver's educational level and their level of practices.

The study concealed that the studied mothers had a highly significant relationship between knowledge and their educational level, occupation, and social class which was supported by **Abd-Elkodoos et al., 2018**, who said that there was a highly statistically significant positive correlation between the educational status of the family caregivers.

According to UNICEF the majority of women in Egypt were illiterate or had primary education and also most of female in Egypt are less educated especially in rural area due to socio-economic and geographical factors, also gender disparities continue to affect access to primary education **(UNICEF, 2016).**

The study revealed that statistically significant relationship statistically significant relationship between mothers practice and their educational level that in contrast with **Roby et al., 2021** who mentioned that no relation between the family caregiver's educational level and their level of practices. Educated mothers frequently have more knowledge and practices compared with illiterate, basic or less educated mothers.

The study showed that there was a statistically significant relationship between mothers' total practice and their age. These results were supported by **Abd-Elkodoos et al., 2018**who mentioned that there was a highly statistically significant negative correlation was found between the total practice scores of the family caregivers and their age.

Regarding the mothers' practices, the current study revealed that more than three quarters had unsatisfactory practices. This result is supported by **Abd-Elkodoos, et al., 2018a** who detected a highly statistically significant negative correlation was found between the total practice scores of the family caregivers and their ages.

Conclusion:

The present study concluded that most of the studied mothers had poor knowledge regarding PKU. Moreover, the majority of them had negative attitude and unsatisfactory practice.

Recommendation:

Based on the results of the present study, the researcher suggested the following recommendations:

- Health education programs and training regarding phenylketonuria should be provided for mothers to improve their knowledge, attitude and practice. Presence of educational kitchen in all clinics for phenylketonuria children to educate mothers how to make healthy meals with low or free phenylalanine.
- Dissemination of health awareness concerning disabilities especially PKU children through the media (radio and television) that invades all homes in the rural and urban areas.
- Furthermore research about health problem, needs and methods of caring PKU children.
- Provide a lot of places for selling any product with low phenylalanine for PKU children at affordable prices.
- Health education programs regarding phenylketonuria should be provided for mothers to improve their knowledge, attitude and practice.

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