Dietary Adherence Regimen Program for Mothers of Children with Phenylketonuria Based on behavior Cognitive learning Theory

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Abstract

Background: phenylketonuria (PKU), cause severe intellectual disability in children if neglected and can lead to utmost predominant effects of mental delay. Dietary compliance depends on people caring for PKU children through modifying their behaviour and knowledge. **Aim**: this study aims to evaluate the dietary adherence regimen program for mothers of children with phenylketonuria based on behavior cognitive learning theory. **Research design**: A quasi-experimental design was used in this study. **Settings:** The study was conducted at the nutrition clinic and Specific Hereditary Outpatient Clinic in Children's Hospital associated to Mansoura University Hospital at Mansoura city, Egypt. **Subjects and method**: the sample comprised of 100 mothers of children with PKU. Purposive sample technique was used to recruit study participants**. Tools**: The researchers used one tool names structured interview sheet: that contain four parts. Part I: demographic characteristic, part II: child medical history, part III: Mothers' knowledge, part IV: grounded on Behavioral cognitive learning theory (pre and post). **Results**: this study displayed statistically significant enhancement in mother's knowledge, behaviour cognitive learning theory items (22.21 ± 6.83 to 26.90 ± 7.19) (60.27±36.67 to 101.02±40.78), respectively pre VS post program. The researchers **concluded that** there were improving in mothers' knowledge, attitude, beliefs, subjective norms, and self-efficacy regarding dietary adherence regimen program of their phenylketonuria children based on behaviour cognitive learning theory. **Recommendations:** continuity of dietary adherence regimen program for mothers of their children affected with phenylketonuria based on behaviour cognitive learning theory ongoing dietetic input is needed to further enhance knowledge of the PKU diet.

Keywords: Behavior cognitive learning theory, Children, Dietary adherence regimen program, Mothers, Phenylketonuria.

Introduction

Phenylketonuria (PKU) is an autosomal dominant metabolic condition caused by diminished phenylalanine (PHE) conversion to tyrosine due to impaired phenylalanine hydroxylase (PAH) activity. Intellectual disability is commonly accompanied by additional indications, including motor abnormalities, mental disorders, inappropriate behaviour, and epilepsy. The most serious result is the poor development of the central nervous system [(1)].

The occurrence of PKU changes international, the mean occurrence is around

1:10,000 children in Europe and approximately 4 cases per 100,000 individual, with a greater rate in some countries as Turkey and Ireland, Scotland, the Yemenite Jewish population, Denmark, France, Estonia, the United Kingdom, Italy, Brazil, Canada, China, and the former Yugoslavia and a very low level in Finland and Africa [(2)].

Moreover, the problem spread in Egypt from late detection and incompetent Newborn Screening (NBS) program and the mothers not concerned with methods of dealing with daily care with children illness. Phenylketonuria prevalence in 2015 revealed that in the Middle East region, induced incidence to population estimated used was 10% in Egypt; 11% in Libya; 14% in Iraq [(3)]. Although, prevalence in Middle East region is very low but the associated burden and adverse effect on children and their families is high consequently.

Dietary adherence regime is the cornerstone of phenylketonuria (PKU) therapy, which is associated with neurocognitive outcomes in childhood. However, adherence to low-phenylalanine diets gradually declines after adolescence, so people with early-treated PKU can achieve normal or near-normal IQ if they maintain good metabolic control. There are four methods of dietary regimen strategy that used to preserve plasma PHE concentrations which sustenance optimum growth, development, and mental functioning although providing a nutritionally complete diet [(4)].

The National Society for PKU (NSPKU) applauded and afterwards advised a lifelong dietary adherence regimen Programme to therapy under the European Rules for PKU, the most current edition, which was issued in 2017. A low- PHE diet is based on four key ideas: Exclusion of foods high in protein and PHE, such as meat, fish, eggs, cheese, bread, flour, pasta, nuts, seeds, and aspartame; measured amounts of PHE from food sources, replacement of the majority of natural protein with an artificial one; and use of very low protein foods [(5)].

Mothers must set a good example In order to effectively manage childhood phenylketonuria, through necessitates everyday effort to address the children's disabilities and to keep a specific diet Programme and rehabilitation; regularly increase their understanding with new foods, maintain constant meal routines, and allocate sufficient time for eating [(6)].

The behavioral cognitive learning theory contends through theorizing the learning process and social behaviour that new behaviour can be acquired by seeing and imitating the actions of others. It advocates learning as a cognitive process that occurs in a social context, can be positive through observation or direct instruction, links beliefs to behaviour, and is intended to be quick, useful, and goal-oriented while also giving people the long-term skills need to stay healthy. Theoretically, an individual's behavioral intentions are influenced by four factors: attitude, self-efficacy, subjective norms, and knowledge. The components of attitude—beliefs, previous experience, and emotional reactivity—serve as rules for encoding pertinent reinforcement contingencies. Rules, on the other hand, act as strong cognitive discriminative stimuli

that significantly affect present and future behaviour [7].

Significance:

Our Egyptian children are burdened by phenylketonuria, which has negative impacts like mental impairment, developmental issues, and behavioral abnormalities. The issue grew in Egypt as a result of inadequate newborn screening (NBS) programmes, late discovery, and moms who were unconcerned with how to treat sick children on a daily basis.

Community nurses play a crucial role in the prevention and control of PKU by educating parents and managing the newborn screening process to ensure healthy outcomes for families. They also offer premarital counseling about relative marriage (endogamy), take advantage of the chance to practice communication skills, and deliver complex information to parents and families. The nurse works together and coordinates with other team members to expedite the referral of a newborn that has been eventually diagnosed with PKU. The nurse also assumes responsibility for correct nutritional management (a diet low in phenylalanine) and participates in the PKU patient's rehabilitation process. Educates parents how to cope with their child's residual disability, make the most of their strengths, and motivates the family to finish the treatment plan and maintain long-term health. [8].

Study aim

This study's aim was to evaluate the dietary adherence regimen program for mothers of children with phenylketonuria based on behavior cognitive learning theory

Research hypothesis:

Providing dietary adherence regimen program for mothers of children with phenylketonuria based on behaviour cognitive learning theory will improve mothers' knowledge, attitude, beliefs, subjective norms and self-efficacy for mothers about their children affected with PKU.

Subjects & Method

Research design:

A quasi-experimental one group pretest-posttest design was used.

Setting:

At Mansoura University's Children's Hospital, specified to nutrition clinic and the specific hereditary outpatient clinic this study was carried out.

Subject and sampling:

The sample comprised of 100 mothers of children with PKU. Purposive sample was used to recruit study participants who attended the previously mentioned setting and available at the time of data collection.

Tools of data collection:

The researchers designing tool as the following after reviewing the recent literature. Structured interview sheet, It includes the following four parts:

Part I: Mothers' socio-demographic data as: mother age, degree of education, current work, and number of living children, children's data as age, and sex.

Part II: Medical history of children as: disease duration, phenylalanine plasma levels, adhere to a low-phenylalanine diet and problems in taking amino acid.

Part III: Structured interview questions was used to assess of mothers' knowledge about

PKU that contain two parts cover general knowledge about PKU and specific knowledge related to nutrition and regimen program as: The disease's definition, causes, symptoms, diagnosis, course of treatment, nutrition, and consequences of adherence regimen program.. It was developed based on **(Witalis, et al., 2017[9].; University of Washington PKU Clinic (2018) [10]**.

Scoring system:

zero score gave to incorrect answer and one score gave to correct answer, the knowledge questions divided into two-part MCQ type that represent 16 questions and true and false types that constitute 20 questions with total score 36 score level divided to three category as the following:

-Good level that represent >75% (27.1-36) of the total scores

-Fair level that represents 50:75 (18-27) of the total scores

-Poor level that represent < 50% (0-17.9) of the total scores

Part IV: Structured interview sheet was grounded Behaviour Cognitive learning Theory adopted from **(Bandura 1986[11].; Ajzen 1991) [12]**. This theory proceeds into account people's subjective norms, self-efficacy and attitudes, the theory describes people's behaviour by taking into consideration their attitudes, self-efficacy, and subjective norms. Attitude composed of 15 belief items, including "consider it is important for my child's health that he/she consistently adheres firmly to the diet," were used to gauge the expected-beliefs build. The attitude construct also took into account the mothers emotional responses to PKU and the diet including "I feel bad when my child eats normal food." The responses to these questions were also rescored in categories, and each of the questions was analyses separately four items were used to assess the emotional reactivity construct as well as three elements served as indicators for the past-experiences construct. 'I strongly disagree' to 'I strongly agree' were the only response options for each topic. A typical example was 'My child continuously adheres well to the diet even if his or her PHE concentrations are from time to time too high'.

A person's opinion that the majority of their "significant others" believe they should behave a certain way is known as a subjective norm. Subjective norms are made up of cues to conform and normative assumptions. The degree to which respondents' spouses, family members, or acquaintances valued the PKU patient's adherence to the diet was used to gauge their normative beliefs which composed of two items. Mothers' self-efficacy was assessed using ten items. Self-efficacy refers to a defendant's belief in his or her ability to act in a particular way. According to a similarity analysis, the answer categories of the items could be seen as continuous.

Scoring system:

One score gave to strongly disagree answer, two score gave to somewhat agree, and three score for strongly agree, the answers to some items were re-scored in categories in posttest than pretest and the items were included separately in the analysis of total score summed by mean and SD.

Procedure

Phase I: Preparatory phase:

1. Administrative Design:

The Mansoura University Dean of the Faculty of Nursing obtained a certified letter to be submitted to the Administrator of the Children's Hospital of Mansoura University, as well as to the Managers of the nutrition clinic and Specific Hereditary Outpatient Clinic.

2. Ethical Considerations:

The Scientific Ethical Committee of the Nursing Faculty at Mansoura University granted its approval (ref. no. p.0421). Additionally, before to data assemblage, each member's signed informed consent was sought. They received guarantees of their identity, confidentiality, and the freedom to take any conclusions from the study at any moment. Respect was shown for morality, philosophy, and beliefs.

Phase II: Operational phase:

1. **Literature review**

It was suggested to review the literatures on the phenylketonuria disease at the national and international levels using academic publications, online searches, and textbooks. The review served as a blueprint for creating the study tools.

Developing the study tools:

The tools were used after reviewing the related recent literature.

Content validity and reliability:

1) To examine the tool parts for clarity, relevance, thoroughness, understandability, applicability, and the necessary revisions, juries composed of five experts in the fields of community health nursing and medicine were convened.

2) Pilot research was conducted on 10 mothers who had PKU-affected children to assess the tool's applicability, viability, and clarity. Mothers and their kids from the pilot study were included in the study sample because the tool did not require any significant modifications.

3) The reliability of the generated instruments was evaluated using the Cronbach's Alpha test through the SPSS Programme version 22 on 10 mothers of PKU-affected children. Reliability testing was completed (0.86).

Field of work:

A formal letter requesting consent to begin the study was approved. In order to obtain support and assistance in the data collection and application of the dietary Programme, this letter incorporated the purpose of the study and a print from the data collection instruments.

Undoubtedly, there were some clarifications. Every meeting lasted between 30 and 45 minutes. Mondays and Wednesdays (both days of the week) from 9 am to 1 pm were used for this investigation. Data was collected around four months ongoing (starting at February and ended in June 2023). Namelessness of mothers‘ responses was guaranteed and secrecy of data was preserved, (25 mothers by researchers in one week that constitute 100 mothers in one month to complete pretest questionnaire and by taking telephone number for the follow up visit provide the program then immediate posttest, the program provided by take five to eight mothers a day to become ten to sixteen mothers throughout the week to complete a hundred mothers within two months and half, as for the rest of the period,

it was for the mothers who did not adhere to the immediate follow-up dates .

Program development:

Initial phase, implementation phase, and evaluation phase were its three phases.

First phase:

The initial phase of this study's development saw the accomplishment of the following objectives :

The researchers met mothers of PKU-affected children who agreed to participate in the study. They explained the purpose of the study and its objectives, which included evaluating the impact of a dietary adherence Programme on the knowledge, attitude, beliefs, and self-efficacy of mothers of PKU-affected children. After ensuring that all information was confidently gathered, the mothers gave their verbal or written consent before the Programme began. In order to establish a relationship of trust and get the mothers' cooperation, the researchers stayed with them .

-Assessment by the previous instruments was done by studying past and recent literature cover many features of the study in books, articles, periodicals, magazines and readings related to the research study.

Implementation phase:

Researchers divided the study group into ten small groups to conduct sessions based on mothers' documentation needs following data collection during the pretest. Such as the main conclusion from pretest results using group discussions and role play to permit everyone clarify her thoughts and prior experiences when dealing with a child's illness, this phase started with the program's implementation. The mothers received the handouts from the researchers, and sessions were used to carry out the programme. The material was covered in twenty weeks, beginning with February and ended in June 2023 that overview of PKU based on behavioral cognitive learning theory that help in behaviour change especially in long term condition or problems as eating disorder through inform mothers about disease, definition, causes, signs and symptoms, methods of prevention, nutritional adherences and treatment; improve towered positive attitude and beliefs also enforced self-efficacy.

Evaluation phase:

Based on the program's success, a post-test was conducted with mothers to gauge the program's impact using the same pre-program tools, and a second reanalysis was conducted to evaluate the program's impact .

Statistical Approach

The statistical package for social sciences (SPSS), version 22, was used to analyses the data. Numbers and percentages were used to represent quantitative data. The demographic and PKU data mean and standard deviations, as well as the results of the t test, Chi-square test, and Pearson correlation coefficient, were recorded. P value was used to indicate that the results of the comparison between the pre- and post-test were statistically significant if less than 0.05.

Results

Table (1): Cites study demographic characteristics of mothers of PKU-affected children's, 58% aged from 25 :< 35 with mean age32.90±6.17, 38% had secondary education, and 66% of them were housewife.

Table (2): Presents study demographic characteristics of children affected with PKU that most affected children are boys aged 11 years and more, regarding educational level 68% in school age. 54% were first ordered in birth and 66% living in rural area.

Table (3): Shows that 38% from studied sample discovered their disease at less than 3 years, 55% diseased for more than one year. 66% not adhere their child to low PKU diet, although 16% monitor child PKU plasma level regularly.

Table (4): This table delineates significant change in mean general and specific knowledge level post versus base line about PKU and its dietary adherence regimen.

Table (5) and figure (1): Demonstrate significant change in total knowledge score post versus preprogram implementation at p value 0.05.

Figure (1): Percentage Distribution of Mothers Regarding PKU Pre / Post Dietary Adherence Program (N=100).

Table (6): Describe significant change in mean behavioral cognitive level post versus base line.

Table (7): Explain significant correlation between total knowledge change and total behavioral cognitive pre and post program implementation.

Table (1): Demographic characteristic of mothers for their children affected with PKU (n=100)

variable	n	%
Age in years		
18:<25	6	6
25:<35	58	58
35:≤45	36	36
mean ± SD	32.90±6.17	
Level of education		
Don't read and write	14	14
Read and write	11	11
primary education	17	17
secondary education	38	38
High education and higher	20	20
Mothers occupation		
Currently employed	34	34
Housewife	66	66

Table (2): Demographic characteristic of children affected with PKU (n=100).

variable	n	%
Age in years		
5:< 8	10	10
8:<11	25	25
11:≤ 15	65	65
mean ± SD	12.16±1.51	
Gender		
boys	63	63
girls	37	37
Level of education		
Preschool (1–5 years)	32	32
School age (6–15 year)	68	68
Birth order		
First	54	54
Middle	33	33
last	13	13
Residence		
Urban	34	34
rural	66	66

Table (3): Distribution of history of children affected with PKU (n=100).

Variable	number
Discovered time	
<3	38
3- <6	26
6- < 9	22
9- ≤15	14
Disease duration	
Recently diagnose	24
One year	55
More than three	21
Medical examinations over the last month	
Once	61
Twice	39
Child adhere to a low- PKU diet	
yes	34
no	66
Monitor child PKU plasma levels regularly	
Yes	31
No	52
Sometimes	16

Table (4): Mean Difference of total knowledge and sub items among mothers of their children affected with PKU pre / post Dietary adherence program (n=100).

variable	Pre program	Post program		
	Mean±SD	**Mean±SD**	**T**	**sig**
General knowledge about PKU	10.31 ± 3.14	12.80 ± 3.29	16.90	.004*
Specific Dietary knowledge	11.90 ± 3.44	14.10 ± 3.71	18.26	.002*
Total knowledge	22.21 ± 6.83	26.90 ± 7.19	23.16	.000*

Table (5): Distribution of mothers' knowledge regarding PKU pre / post dietary adherence program (n=100).

variable	Mothers' knowledge pre and post PKU dietary program				χ^2	P
	Pre-program		Post- program			
	No.	%	No.	%		
Total knowledge levels:						
Poor	58	58	18	18		
Average	31	31	37	37		
Good	11	11	45	45		
Total knowledge mean scores preprogram: Mean ± SD	22.21 ± 6.83				6.03	0.018*
Mean change of scores of total knowledge post program: Mean ± SD	26.90 ± 7.19					

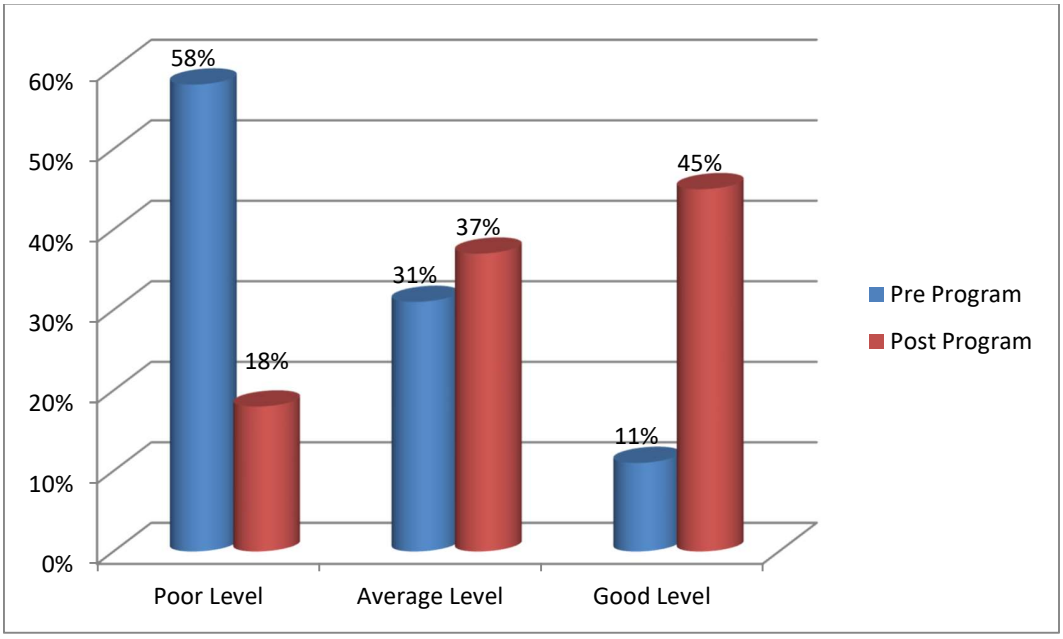

Figure (1): Percentage distribution of mothers regarding PKU pre / post dietary adherence program (n=100).

Table (6): Mean Difference of total Behavioral Cognitive learning Theory and sub Items among mothers of their children affected with PKU pre / post dietary adherence program (n=100).

behavioral cognitive	Pre program	Post program		
	Mean±SD	Mean±SD	t	sig
attitude				
Beliefs (15-45)	25.8±7.34	43.91±6.93	16.90	.004*
Past experience (3-9)	4.01±5.16	8.98±2.19	18.26	.002*
Emotional reactivity (4-16)	7.4273±2.78086	14.1091±3.71297	26.40	.000*
Subjective norms(2-6)	2.91±1.42	5.24±1.93	20.49	.001*
Self-efficacy (10-30)	20.09±6.77	28.79±7.77	19.37	.003*
Total behavioral cognitive (34-102)	60.27±36.67	101.02±40.78	23.16	.000*

Table (7): Correlation between changes of knowledge, and Behavior Cognitive learning Theory regarding PKU pre/Post Dietary Adherence Program (N=100)

variable	Change of scores of total knowledge, and total behavioral cognitive			
	Pre dietary adherence program total mothers knowledge		post dietary adherence program total mothers knowledge	
	r	p	r	p
Total behavioral cognitive	.998	0.000*	.668	0.000*

Discussion:

In nations without a newborn screening programme, PKU is main greatest causes of mental retardation and, if untreated, is linked to severe intellectual disability in children. PKU affects 1 in 23,930 live births worldwide. The mainstays of the diet- based treatment for PKU are low-phenylalanine diet and the monitoring of phenylalanine levels. Challenging for diet regime adherence are limited access to low-protein modified foods and careers' inadequate knowledge of and improper practices regarding the PKU diet. Moreover, children with PKU may lack micronutrients[13]. **(Zamani, et al., 2021).**

The recent study found that, nearly more than half of studied mothers had poor knowledge about PKU preprogram this may be due to majority of the studied sample had low educational level and nearly three quarter experience the disease with affected

child to about one year and these results agree with **Elsayed.et, al (2020)** [14], In Cairo, Egypt, who assessed mothers care toward their children having Phenylketonuria" and reported that more than half of mothers had poor knowledge about PKU and **Abd-Elkodoos. et al., (2013[15],** study at Abo El Reesh Hospital and reported that the majority of children received poor nutritional control and care notwithstanding the fact that the family members had low levels of knowledge.

As reported by Ozel, et al.,(2008[16].), Turkey, who study in Turkish children with phenylketonuria" and shows that just 3.5% of mothers had poor disease knowledge (mean score of 50% for disease knowledge), but 34% had poor dietary knowledge and 84.7% had poor understanding of the phenylalanine exchange system.

In the same line Öztürk, etal., 2022[17] who study " parents´ knowledge regarding phenylketonuria and affecting factors" and revealed that parents' knowledge regarding the disease condition and food therapy were inadequate.

Accordance with **Witalis, et al., 2017[9]**, who study "Phenylketonuria patients' and their parents 'knowledge and attitudes to the daily diet -multi-center study" and showed that only 27% of parents (n = 41) understood the PHE content in at least three out of four examined food categories, and only 45% (n = 74) of PKU patients and parents were aware of daily PHE intake guidelines.

Therefore, improving diet PKU adherence could benefit from focusing on the variables suggested in the Integrative Model of Behavioral Prediction (IMBP) theoretical framework. It makes the same assumption as Behaviour Cognitive Learning Theory that intention is the primary predictor of behaviour. This theory proposes that self-efficacy, subjective norms, attitude, and supportive environments and skills, which serve as actual control variables, regulate the link between intention and behaviour. **(Dai and Harrington, 2021)** [18].

Concerning the effect of program on the level of behavioral cognitive learning theory and total knowledge level pre and post program the present study reported that significant improvement in mothers' total level of behavioral cognitive learning theory and sub items, additionally, shows significant improvement in mothers' knowledge, these results agree with **Fouad, and Abd Elmoneem (2017)** [19]. who implement Intervention Program for Family Caregivers Having Children with Phenylketonuria and reported that the total satisfactory knowledge for family caregivers varied in a way that was statistically significant with baseline preprogram.

In the same line, "An intervention program by **Abd-Elkodoos, et al 2018[20].,** for family caregivers of children with phenylketonuria' knowledge and practices" who reported that the program considerably improved the family caregivers' knowledge and practices regarding caring for their phenylketonuria children.

Similarly, to **Ozel, et al. (2008)** [16] who claimed that there were statistically significant differences in family caregivers' knowledge regarding the meaning of PKU,

disease causes, its signs and symptoms, the various available treatments, prevention of potential complications of the disease, and warning signs to see a doctor, there were statistically significant differences in their knowledge post-test compared to pre-test. Following the implementation of the nursing Programme, these differences demonstrated a substantial improvement in the satisfaction of all family caregiver's' knowledge (X2 = 28.564 at P 0.001).

Congruent with Rahgoi, et al.,2019[21]. who study the " Burden of Care in Mothers of children affected with Phenylketonuria" empowerment program", and reported that in the intervention group, mothers of children with PKU had a mean score of total care burden and its dimensions that was considerably lower in the post-test than in the pre-test (41.20 5.04 vs. 58.24 3.96; P 0.001).

These results agree with **Fouad, and Abd Elmoneem (2017) [19]**. **and Zamani, et.al 2021[13]**, whom a pilot randomized study, indicated that empowering caregivers of patients, building strong social networks, giving positive social supports, and facilitating access to specialized foods may all help regulate and control PKU level.

The current study reports that after adherence programme implementation, mothers' attitudes, subjective norms, and levels of self-efficacy and supporting beliefs, which are sub-items of the behavioral cognitive learning theory, were considerably altered. Additionally, that agree with prior research demonstrated the significance of psychological supports, such as support groups, in enhancing the children's compliance with diet and control of phenylalanine, as reported by **Medford, et al 2017[22]., who study** treatment adherence and psychological wellbeing in maternal careers of children with phenylketonuria **"and with Fishbein, & Ajzen, 2011[23]**, who study, "Predicting and changing behavior: The reasoned action approach and Intention to behavior: Using the integrative model of behavioral prediction" .

The current study shows that there were statistically significant relationships between the mothers' overall cognitive behaviour and their overall knowledge towards caring for their PKU-affected children before and after the Programme. This finding is consistent with **Ali's 2008[24]** study, which found that when parents are given a basic understanding of their child's condition, developmental prognosis, and available treatment options, it helps them adopt healthy behaviour and can help them stop engaging in unhealthy ones. Additionally, this viewpoint is consistent with **a study by Macdonald et al., 2012[25]** who study parent education and knowledge can affect blood phenylalanine control in Phenylketonuria and believed that dietary knowledge is an essential factor on dietary compliance**. Moreover, Ashe, et al (2019) [26]**, who study "Psychiatric and Cognitive Aspects of Phenylketonuria: The limitations of diet and promise of new treatments" and documented dietary advice put the emphasis on continuing dietary treatment for the rest of one's life in order to maximize psychiatric and cognitive outcomes.

Conclusion: The study concluded that the dietary adherence regimen program based on behavioral cognitive learning theory had significantly improved the mothers' knowledge, attitude, beliefs, subjective norms and self-efficacy about the care of their children with phenylketonuria.

Recommendations: based in research hypothesis this study recommended:

- Publication and dissemination of the dietary adherence program for mothers of children with phenylketonuria in all children hospital and family health centers to raise their awareness about the disease management to prevent its complications.
- Replicate in another setting and large sample for generalization.
- Create a phenylketonuria education programme for mothers to give them the knowledge and skills they need to care for their children at home.
- Give out booklets and brochure to mothers to aid in their retention of knowledge for future review.
- Mothers were periodically educated on the illness and nutritional therapies to raise their level of understanding.

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