Assessment of Parents' Performance about Nutritional Management of Children with Phenylketonuria

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Abstract
Phenylketonuria also identified as (phenylalanine hydroxylase deficiency) is an autosomal falling syndrome of phenylalanine metabolic rate, in which particularly high phenylalanine focusses cause mind dysfunction. If crude, these mind dysfunction consequences in severe knowledgeable disability, epilepsy and behavioral difficulties. Purpose: To Assess Parents' Performance according to their assessment of nutritional management; to Identify the association between parents' performance about nutritional management and their demographic variables of children (age, gender, and parents' Socioeconomic Status). Methods: A study was conducted in the following Ibn Al-Baladi for Maternity and Pediatric Hospital (Metabolic disorders and neonatal screening consultants). A non-experimental design; Data was collected after the approval of parents to involve themselves and their children in the study. data are analyzed using Statistical Data Analysis. an instrument was built for parents' nutritional management for their children diagnosed with phenylketonuria to assess the parents' performance. Results: Outcomes presented that 85.7% of parents are showing moderate performance about of nutritional management for children with phenylketonuria during pre-test time, while they show very good level of performance during the post-test time 1 and post-test time 2 (88.6%). Conclusion: Overall Assessment of Parents' Performance showing moderate performance about nutritional management, their performance level is increased to good during the post-test time.

Keywords: Assessment, Parents' Performance, Nutritional Management, Phenylketonuria.

INTRODUCTION

Phenylketonuria also identified as (phenylalanine hydroxylase deficiency) is an autosomal falling syndrome of phenylalanine metabolic rate, in which particularly high phenylalanine focusses cause mind dysfunction. If crude, these mind dysfunction consequences in severe knowledgeable disability, epilepsy and behavioral difficulties [1,2].There are other studies, when apply nutrition management program for any disorders or diseases the result become positive outcome, such as The food instruction should be to concentrate inside substance misuse treatment service area, which can recover treatment consequences and propose nutritionists to be portion of treatment groups in drug misuse units in order to mix nutrition service area in the therapy process, The study focused on nutrition for children with phenylketonuria to prevent mental and intellectual problems throughout life span[3,4,20]. Nutritional treatment may be linked with an increased danger of obesity, in phenylketonuria. The initial studies describe a bent for overheavy in PKU, but then not all new publications approve this, though there stand an increasing amount of studies describing bigger fatness in female clients with PKU. Near is little information describing the metabolic significances of obesity in phenylketonuria [5,6].

MATERIALS AND METHODS

A non-experimental design, the study was conducted on parents of phenylketonuria child in Baghdad city from the period of 3ed June to 5th October 2022; in the following hospital: Ibn Al-Baladi for Maternity and Pediatric Hospital (Metabolic disorders and neonatal screening consultants). Which are located in Baghdad Al-Rusafa side; A Non-probability sample (convenience) of 35 PKU children with 35 of their parents (father and mother) was selected Purposively from the hospital that are select for the study According to the following criteria. Inclusion Criteria for the sample: Children with PKU, according to the diagnosis of pediatrician, consultant of neonatal screening and metabolic diseases, and Parents whose PKU child is between (0 -12) years old; exclusion criteria for the sample: Secondary caregivers (grand father or mother, son, daughter, sibling, and other), and Respondent who did not fill the questionnaire

completely or refuse to answer. A questionnaire is constructed for the purpose of the study throughout a review of relevant literature; It composed of two parts: It includes two sections: first part are Demographic characteristics concerning child such as: age, gender and "Socioeconomic Status" with significant comparison [7], and second part is Assessment of Parents' Performance about Nutritional Management for Children with Phenylketonuria; Describes how parents following a healthy nutritional program for children with phenylketonuria, and the benefit of this program, and it contains (12) items. It is recorded according to the likert standards of 2 points: (1 = known, 0= unknown). The reliability analysis determined by using Cronbach`s alpha coefficient which shows very good evaluation for knowledge scale; the findings mean that the questionnaires had adequate level of internal consistency and equivalence measurability. The data was collected after the approval of the directors of the hospitals and the acceptance of parents to involve themselves and their children in the study, the interview was for parents and children in the same hospital, the questionnaire was completed by parents. The following statistical data analysis approaches were used application of the statistical package (SPSS) ver. (26.0). The Scientific Study Ethical Commission at College of Nursing, University of Baghdad, has accepted the research to be conduct. All participants have taken on a consent formula to present their agreement in place of such membership and keep their human rights.

RESULTS

This chapter presents the descriptive analysis of the sample related to socio-demographic characteristics of children with phenylketonuria and their parents; and describes the level of parents' performance about nutritional management of their children with phenylketonuria. This chapter also describes the relationship between parents' performance and their children variables.

Table 1
Distribution of children and Parents according to their Socio-demographic Characteristics

No	Characteristics		F	%
1	**Child Gender**	Male	19	54.3
		Female	16	45.7
		Total	**35**	**100**
2	**Child Age (year)**	Less than 3	8	22.9
	M±SD= 4.4±1.8	3.1 – 5	21	60
		5.1 – 7	2	5.7
		7.1 – 9	4	11.4
		Total	**35**	**100**
3	**Socioeconomic status**	Low	11	31.4
		Moderate	15	42.9
		High	9	25.7
		Total	**35**	**100**

No: Number, f: Frequency, %: Percentage, M: Mean, SD: Standard deviation

This table shows that more than half of children are males (54.3%) while remaining are females. The average age for children is refers to 4.4±1.8 years in which 60% of them are with age group of 3.1 – 5 years. And show moderate socioeconomic status (42.9%).

Table 2
Assessment of Parents' Performance about Nutritional Management

Assessment	Pre-test				Post-test 1				Post-test 2			
	f	%	M	SD	F	%	M	SD	F	%	M	SD
Poor	1	2.9	12.86	2.932	0	0	20.23	2.522	0	0	20.23	2.522
Moderate	30	85.7			4	11.4			4	11.4		

Good	4	11.4			31	88.6			31	88.6		
Total	**35**	**100**			**35**	**100**			**35**	**100**		

f: Frequency, %: Percentage, M: Mean, SD Standard deviation
Poor= 0 – 8, Moderate= 8.1– 16, Good=16.1 – 24

This table indicates that 85.7% of parents are showing moderate knowledge about application of nutritional management for children with phenylketonuria during pre-test time, while they show good level of knowledge during the post-test time 1 (88.6%) and post-test time 2 (88.6%).

Table 3
Assessment of Items related to Parents' Performance about Nutritional Management for Children with Phenylketonuria

List	Items	Pre-test		Post-test1		Post-test2	
		M	**Asses.**	**M**	**Asses.**	**M**	**Asses.**
1	Nutrition program of phenylketonuria patients is great importance and contribution to the improvement of the child's health condition.	.40	M	.89	Good	.89	Good
2	Children with the disease need to obtain essential nutrients through special dietary supplements due to the limited diet.	.60	M	.80	Good	.80	Good
3	Therapeutic milk is considered as a substitute for breast milk, according to the names (PKU, Phenex Phenylfree).	.71	M	.89	Good	.89	Good
4	Therapeutic milk (phenylalanine-free) is considered a treatment for children as an alternative to breast milk or general alternative milk.	.57	M	.83	Good	.83	Good
5	Infant is given only two feedings, a mixture of breast milk or artificial milk with therapeutic milk.	.63	M	.77	Good	.77	Good
6	Infant (1 day-6 months) is given milk No. (1) of Phenyl Free-1	.66	M	.94	Good	.94	Good
7	Child from the age of (2 years onwards) is given milk number (3) of (PKU3, phenex-3, phenylfree-3).	.40	M	.86	Good	.86	Good
8	Sugars burn the phenyl in the blood, so the patient feels better.	.57	M	.83	Good	.83	Good
9	Meat of all kinds, white and red, such as beef, sheep, chicken and fish is considered a prohibited food for all ages.	.57	M	.77	Good	.80	Good
10	Fats are harmless foods for phenylalanine patients due to their low protein content. The fats are in butter, olive oil, and ghee.	.34	M	.89	Good	.86	Good
11	Avoid all dairy products and foods made from dairy products such as cheese, yogurt, ice cream and high-fat milk.	.57	M	.91	Good	.91	Good
12	Yolk is given from one day to the next and up to.	.49	M	.86	Good	.83	Good

f: Frequency, %: Percentage, Asses: Assessment, (Mean: Poor= 0-0.33, Moderate= 0.34-0.67, Good= 0.68-1)

This table presents the items related to Parents' Performance about Nutritional Management for Children with Phenylketonuria; the mean scores for item during the pre-test time indicate moderate level of knowledge as seen among all items, while during the post-test 1 and 2, the parents' knowledge increased to good among all items as indicated by mean scores.

Table 4
Independent Sample Test for Parents' Performance with respect to Child's Gender (N=35)

Performance	Gender	M	SD	t	Df	p≤ 0.05	Sig
Overall performance	Male	26.32	5.313	.520	33	.607	N.S
	Female	25.50	3.633				

M: Mean, SD: Standard deviation, t: t-test, df: Degree of freedom, Sig: Significance, p: Probability value, N.S: Not significant, S: Significant, H.S: High significant

This table shows that there is no significant difference in parents' performance with regard to child's gender.

Table 5
Analysis of Variance for Parents' Performance with respect to Child's Age (N=35)

Performance / Age	Source of variance	Sum of Squares	df	Mean Square	F	Sig.
Overall performance	Between Groups	2.261	3	.754	.033	.992
	Within Groups	709.625	31	22.891		
	Total	711.886	34			

df: Degree of freedom, F: F-statistic, Sig: Significance

This table reveals that there is no significant difference in parents' performance with regard to child's age group.

Table 6
Analysis of variance for Parents' Performances with regard to Socioeconomic Status (N=35)

Performance / SES	Source of variance	Sum of Squares	df	Mean Square	F	Sig.
Overall performance	Between Groups	4.063	2	2.032	.092	.912
	Within Groups	707.822	32	22.119		
	Total	711.886	34			

df: Degree of freedom, F: F-statistic, Sig: Significance

This table shows that there is no significant difference in parents' performance with regard to their socioeconomic status.

DISCUSSION

Investigation of such parameters presents that greatest of the phenylketonuria children are preschool age males and one third of them is moderate socioeconomic status (Table 1). The demographic characteristic of phenylketonuria children in (Table 1) show that the age of children ranged from 1 to 9 years with mean= 4.4. This result was difference than that described by Bosch and his colleagues (2015) who establish that the children's reaching among 9and 11years with a mean= 9.8. The differences in age between the current study and the study of Bosch and his colleagues (2015) may stay due to the choice criteria of this studies which involves of children age 1-9years' old [8].

The results also show that 54.3% of phenylketonuria children were males and 45.7% were females' (Table,1). This gender distribution is nearly consistent with Bosch and his colleagues (2015) who found that their sample included 43% males and 54 % females. The CDC (2020) stated that PKU is the same for males and females [8]. The researcher believes that this could be because of genetic differences between the sexes but the stay the same morbidities and nearly equal. however, our understanding is far from complete, and this will remain the case until know more about the causes of PKU.

Results of this Table (1) indicate that most of the parents have moderate socioeconomic status (42. 9%).This result was congruent with van Spronsen and his colleagues (2021) who found Socio-economic Status was moderate at (53%) [9]. This result disagrees with Hussein, & Aziz, (2016). who reported most of the mothers (60%) are making low socioeconomic status [10]. This result was supported by Saeed, & AL-Mosawi, (2020). who found the economic situation shows (76.7%) 23 nurses are moderate [11]. The researcher believes that the socio-economic status is moderate in more than study, not low and not high level, an indication of the burden on the parents of children with PKU.

The results in (Table, 2) indicate that parents are showing moderate performance about application of nutritional management for children with phenylketonuria during pre-test time, while they show very good level of knowledge during the post-test time 1 and post-test time 2.

This result was congruent with Rocha and his colleagues (2019), which shows the level of applied the nutritional status and program toward PKU children by parents are moderate and suitable for the sample size [12]. This result is supported by Jameson and Remmington (2020), who found the parents of PKU child follow up low protein nutrition protocol with better nutrition indicators (higher fruit and vegetable consumption) was high level of practices. Furthermore, encouraged low protein eating consumption for PKU children. Still and indeed logical to use low protein diet guidance [13]. This result was congruent with Atshan, & Aziz, (2022). Who show that there is high significant difference in overall main domains relating parents' knowledge of beta-thalassemia major between pre and post-tests 1&2 in the study group [14]. These results agree at over all with Rajih, & mohammed, (2020). who found totally the study illustration responses at the pretest are just knowledge with a numerical mean of scores (1.342). Additional than the posttest, the board exemplifies the (100%) of the research sample have great knowledge at the mean of scores, (1.879) [19]. This result is disagreeing with Van Calcar and his colleagues (2020) who found weak performance of parents with PKU children in overall, must have parents Counsel and their caregivers to provide frequent, high carbohydrate feedings (glucose polymers, or simple or complex carbohydrates) and strive to achieve usual energy intake to prevent catabolism [15].

Regarding (Table,4). Who have studied result show no significant difference relation between the gender of Phenylketonuria child (p-value, 0.607) and parents' performance main sections. These results disagree with Evans and his colleagues (2022) who found highly significant relationship between the gender of Phenylketonuria child (p-value, 0.001) and dietary patterns management at overall. This result supported by Daly et al., (2020), who have studied PKU children non-significant relation between the gender of PKU child and Dietary Intake [16,17]. Related (Table,5), concerning age, the results show there is no significant difference in parents' performance with child's age group (p-value, 0.992). these results agree with Evans and his colleagues (2022), who have studied a dietary patterns management, who found no significant relation between age of PKU child and dietary patterns management [16]. this result supported by reported to be significant differences between pre and posttest of the study sample in each of the study's major domains at P. value equal to 0.000 of nurses" Performances toward Personal hygiene [21].

Concerning (Table, 6) socioeconomic status, the results show that there is no significant difference in parents' performance with regard to their socioeconomic status (p-value, 0.912). This results disagreeing with Nogueira et al., (2021), who found significant relation between family socioeconomic status (p-value, 0.010) and parents' metabolic control [18].

CONCLUSION

The present study Conclusion that: The study findings demonstrate that more than half of children are males while remaining are females; Overall Assessment of Parents' Performance about Nutritional Management for Children with Phenylketonuria showing moderate performance about nutritional management during pre-test time, their performance level is increased to good during the post-test time 1 and 2 as seen among of them; phenylketonuria children's development is influenced by Parents 'Performance about Nutritional Management.

Based on the early stated conclusion, the study recommends that: Parents of phenylketonuria children should be very well-oriented and greatly aware about the importance of Nutritional Management. So, spreading the program on all consultants of neonatal screening; Phenylketonuria children should be screened periodically for growth and development defect monitoring; Families of phenylketonuria

children should be supported in order to help them dealing effectively with such public health problem; Further research studies can be carried out with respect to large sample size and wide-range variables, particularly, nationwide studies.

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