

Effect of nursing intervention on adjustment patterns of children suffering from phenylketonuria.

Manar Fathi Hassan^{1*}, Wafaa El-Sayed Ouda², Safaa Salah Ismail³

¹Department of Pediatric Nursing, Helwan University, Cairo, Egypt

²Department of Pediatric Nursing, Ain Shams University, Cairo, Egypt

Received: 01 November, 2022, Manuscript No. AAJCP-22-79780; **Editor assigned:** 03 November, 2022, Pre QC No. AAJCP-22-79780(PQ);

Reviewed: 17 November, 2022, QC No. AAJCP-22-79780; **Revised:** 23 November, 2022, Manuscript No. AAJCP-22-79780(R); **Published:** 30 November, 2022, DOI:10.35841/0971-9032.26.11.1700-1710.

Abstract

Background: Phenylketonuria is an inherited disorder that increases the levels of a substance called phenylalanine in the blood.

Aim: Improve adjustment patterns of children suffering from phenylketonuria.

Methodology: This study was conducted at genetic clinic in outpatient department belonging/ children's hospital affiliated to Ain Shams university hospitals. A quasi experimental research design was utilized.

A purposive sample of 60 children accompanied by their mothers at the previously mentioned setting over a period of 6 months and satisfying the inclusive criteria of the study.

Three tools were utilized in this study pre/post nursing intervention. The first tool was a structured questionnaire sheet to assess children's knowledge regarding phenylketonuria, the second tool was psychometric assessment to assess psychological problems of children with phenylketonuria; the third tool was adjustment patterns scale of children with phenylketonuria.

Results: The study findings revealed that there was statistical significant difference between the studied children's knowledge pre and post nursing intervention implementation and there was statistical significance difference regarding to total adjustment patterns of the studied children pre and post nursing intervention implementation. Also, there was an extremely statistical significant difference and positive correlation between children's total knowledge and total adjustment patterns pre and post nursing intervention implementation.

Conclusion: There was there was a positive effect of the nursing intervention on the adjustment patterns of children suffering from phenylketonuria.

Recommendations: Continuous health educational programs based on their actual need assessment about adjustment patterns should be provided for children suffering from phenylketonuria to cope effectively with their disease.

Keywords: Adjustment Patterns, Children, Intervention, Program, Phenylketonuria, Nursing.

Accepted on 15 November, 2022

Introduction

Phenylketonuria is an inherited metabolic disorder resulting from mutations in the PAH gene. These mutations result in an impaired ability of the enzyme to metabolize Phenylalanine (Phe), to Tyrosine (Tyr), leading to the accumulation of Phe in blood and tissues. Allelic variation in the PKU children results in a broad spectrum of severity of PAH dysfunction and thus a wide range of clinical manifestations [1]. The prevalence of PKU varies worldwide. In Europe, the mean prevalence is approximately 1:10,000 newborns with a higher rate in some countries such as Ireland and Turkey and a very low rate in Finland [2].

Untreated PKU results in the slow insidious loss of neurocognitive skills resulting in permanent cognitive impairment as the child grows. Classical signs include eczema like skin rash, excessive restlessness and a "musty" or "mousy" odor of the body,

urine and perspiration due to phenyl acetate accumulation. In addition, children affected with PKU have a lighter coloration of skin, hair and eyes. Developmental problems, irritable behavior, gait disturbances, psychiatric symptoms and impaired cognition become clinically present with increasing toxic accumulation within the body and brain [3].

Children with PKU have behavioral problems including hyperactivity, stereotypy and anxiety. Children with PKU may also show varying combination of denial, depression, lack of self-confidence, lack of independence and limitations of social communication. Therefore, it is necessary for pediatric nurses to know how children with PKU manage their activities of daily living, medical regimen and social relationships. For that, the nurse through nursing intervention can provide support to phenylketonuric children in a number of adjustment patterns [4].

Pediatric nurses play a crucial role in assisting phenylketonuric children to deal with perceived stressors, changes or threats which interfere with meeting life demands and roles through educating them other ways of adjustment such as seeking information, reprioritizing needs and roles, lowering expectations, making compromises oneself to other planning activities to conserve energy, taking things one step at a time, listening to one's body and using self-talk for encouragement. The nurses can implement the adjustment patterns and explore methods for improving the children's adjustment abilities [5].

Aim of the study

This study aimed to improve adjustment patterns of children suffering from phenylketonuria.

Research hypothesis

The nursing intervention will positively affect the adjustment patterns of children suffering from phenylketonuria.

Methodology

Research design

A quasi experimental design was utilized to achieve the aim of this study.

Research setting

The study was conducted at genetic clinic in outpatient department belonging to children's hospital affiliated to Ain Shams university hospitals.

Study subjects

A purposive sample was composed of 60 children accompanied by their mothers in the previously mentioned setting over a period of 6 months and satisfying the following:

Inclusion criteria

Children with confirmed diagnosis of phenylketonuria in age group of 6 to 12 years regardless their gender, residence and level of education.

Exclusion criteria

Children with either chronic medical or mental health problem.

Tools of data collection

Data were collected through using the following tools pre and post nursing intervention.

Tool I: Structured questionnaire sheet: It was designed by the researcher after reviewing the current available literature and it was written in simple Arabic language to assess the following:

- Characteristics of the children which include; age, gender, birth order, residence and history of the disease.
- Characteristics of parents of phenylketonuric children which include: age, level of education, occupation, monthly income and family history of the disease.
- Knowledge of children regarding phenylketonuria which include; definition, causes, signs and symptoms, treatment,

complications, prevention of phenylketonuria, prevention of complications and nutrition.

Tool II: Psychometric assessment: It was used to assess psychological problems of children with phenylketonuria through.

Children Depression Inventory (CDI): Children Depression Inventory (CDI) was originally prepared by Kovacs, and modified by Abdel-Fattah, to assess the degree severity of depressive symptoms. It fit children from age 6-18 years. It consisted of 27 statements such as grief, failure feelings, wrong behavior, optimism, suicidal thoughts and social problems [6].

Scoring system: each statement was answered by always, sometimes or never. The children were scored two if the answer is "always", one if the answer is "sometimes" and zero if the answer is "never". The total score ranged from 0:54. Regarding to severity of depression symptoms, score 0 referred to no depression symptoms, score 1:<18 referred to mild depression symptoms, score 18:<36 referred to moderate depression symptoms and score 36: ≤ 54 referred to severe depression symptoms [7,8].

Children Manifest Anxiety Scale (CMAS): Children Manifest Anxiety Scale (CMAS) by Castaneda et al. [9] that was modified by Abdel-Hamid et al. [2] to assess the degree of severity of anxiety symptoms of children. The anxiety scale consisted of 36 statements. It measured all the symptoms of anxiety namely somatic, emotional, motor and social symptoms.

Scoring system: each statement was answered by "yes" or "no". The children were scored zero if the answer is "no" and one if the answer is "yes". The total score ranged from 0:36. Regarding to severity of anxiety symptoms, score 0 referred to no anxiety symptoms, score 1:<12 referred to mild anxiety symptoms, score 12:<24 referred to moderate anxiety symptoms and score 24: ≤ 36 referred to severe anxiety symptoms.

Children Loneliness Scale (CLA): It was originally prepared by Rasel and modified by Al-Behery to assess children's feeling of loneliness. It also modified by the researcher to suit nature of the study. The loneliness scale consisted of 20 statements [4,5].

Scoring system: Each statement was answered by always, sometimes or never. The children were scored two if the answer is "always", one if the answer is "sometimes" and zero if the answer is "never". The total score ranged from 0:40. Regarding to severity of loneliness symptoms, score 0 referred to no loneliness symptoms, score 1:<13 referred to mild loneliness symptoms, score 13:<26 referred to moderate loneliness symptoms and score 26: ≤ 40 referred to severe loneliness symptoms.

Self-Esteem Inventory (SEI): Self-Esteem Inventory (SEI) was developed by Smith, and modified by El-Dosoki et al. [6], to measure self-esteem of children. It evaluates attitudes toward the self in social, academic, family and personal areas of experiences and consisted of 20 statements.

Scoring system: Each statement was answered by always, sometimes or never. The children were scored two if the answer is "always", one if the answer is "sometimes" and zero if the answer is "never". The total score ranged from 0:40. According

to the given responses of the studied children, self-esteem was categorized into: score <13 referred to low self-esteem, score 13:<26 referred to moderate self-esteem and score 26: ≤ 40 referred to high self-esteem.

Tool III: Adjustment patterns of children with phenylketonuria: Adjustment patterns of the children with phenylketonuria concerned with positive and negative adjustment patterns of children toward their home, school, social and medical compliance. Adjustment patterns constructed by the researcher based on relevant studies.

Scoring system: Each statement was answered by always, sometimes or never. The children were scored two if the answer is “always”, one if the answer is “sometimes” and zero if the answer is “never”.

Content validity and reliability

The revision of the tools for face, content validity, its clarity, relevance, comprehensiveness, understanding and applicability was done by a panel of 3 experts in the field of pediatric nursing then the necessary modifications were done accordingly. Internal consistency and reliability were measured by using Cornbrash's alpha-coefficient test.

Pilot study

A pilot study was carried out on 10% of children with phenylketonuria attending in the previously mentioned setting to test the applicability, clarity and efficiency of the tools. Then the necessary modifications of the tools were done according to the results of pilot study in the form remediation of some items. The pilot study had also served to estimate the time needed for each subject to fill in the study tools. Children involved in pilot study were excluded later from the main study sample.

Field work

The actual field work was carried out in the first week of February 2020 up to the end of July 2020 for data collection. The researcher was available in the above previously mentioned setting twice weekly (Monday and Wednesday) from 9 am to 2 pm. The researcher started by introducing herself to children and their mothers and gave them a brief idea about the study aim and its expected outcomes. Tools were filled by interviewing each phenylketonuric child and accompanied mother individually for 20 to 30 minutes to gather the necessary data of the study. The program was carried out at the previously mentioned setting. The nursing intervention had been sequenced through 10 sessions. Subjects were divided into subgroups according to children's age and the nursing intervention was implemented for each group separately (2 days/week). The program lasted for 5 months in addition to one month for pretest and posttest. The duration of each session was ranged from 30-45 minutes. Different teaching methods were used such as lectures, small group discussion and role play. Upon the completion of the nursing intervention, the post test was done to evaluate the outcomes of the indicators of nursing intervention success.

Administrative design

An official permission to conduct the study and implement the nursing intervention was obtained from the hospital administrators of the study setting through a formal letter that

was issued from the dean of the faculty of nursing, Helwan University.

Statistical design

The collected data were organized, reviewed, coded and tabulated. Statistical analysis was done by computer with Statistical Package for Social Science (SPSS) version 20 as used to estimate the statistical significance difference between variables of the study. Data were presented using descriptive statistics in the form of frequencies and percentages. Quantitative data were presented in the form of $X \pm SD$. Qualitative variables were compared using chi-square test (X^2) to compare between two qualitative variables. Statistical significance was considered at p -value<0.05. Highly statistical significance was considered at p -value<0.001. Extremely statistical significance was considered at p -value<0.0001.

Results

Table 1 showed characteristics of the studied children. It revealed that three quarters of the studied children (75%) were aged 10: ≤ 12 years ($X \pm SD$; 10.16 ± 1.51 years) and more than half of them (55%) were males. Moreover, most of the studied children (80%) were in primary school and almost two thirds of them (65%) were ranked as the first order. It was cleared that more than three quarters of the studied children (76.7%) came from rural residence.

Table 2 showed history of children's disease; it was found that about half of the studied children (46.7%) were diagnosed with phenylketonuria in age less than 3 years and half of them (50%) discovered the disease by investigations. Regarding the number of follow per month, it was revealed that more than two thirds of the studied children (68.3%) had follow up once per month.

Table 3 showed parents' characteristics. It clarified that; almost two thirds of the studied mothers (63.3%) were aged 25:<35 years ($X \pm SD$; 32.90 ± 6.17 years). On the other hand, it was cleared that more than half of the studied fathers' age (61.7%) were 35:<45 years ($X \pm SD$; 36.85 ± 6.59 years). Regarding to the educational level half of the studied mothers (50%) and more than half of the studied fathers were having technical education. Furthermore, more than three quarters of the studied mothers (76.7%) were not working, while 86.7% of the studied fathers were working.

Table 4 revealed that there was statistical significant difference between the studied children's knowledge pre and post nursing intervention implementation. It was noticed that three quarters of the studied children (75%) had unsatisfactory knowledge regarding phenylketonuria pre nursing intervention implementation. On the other hand, it was observed that most of them (81.7%) had satisfactory knowledge regarding phenylketonuria post nursing intervention implementation.

Table 5 showed that there was extremely statistical significant difference between the studied children's level of depression pre and post nursing intervention implementation, where more than half of the studied children (58.3%) had level of moderate depression pre nursing intervention implementation, while post nursing intervention implementation more than three quarters of them (78.3%) had mild level of depression.

Variables	Number (No)	Percentage (%)
Age in years		
6:<8	5	8.3
8:<10	10	16.7
10: ≤ 12	45	75
X ± SD	10.16 ± 1.51	Range
Gender		
Male	33	55
Female	27	45
Level of education		
Not yet enrolled	2	3.3
Primary	48	80
Preparatory	10	16.7
Birth order		
First	39	65
Middle	18	30
Last	3	5
Residence		
Urban	14	23.3
Rural	46	76.7

Table 1. Number and percentage distribution of the studied children according to their characteristics (n=60).

Variables	No.	%
Child's age at diagnosis of the disease in years		
<3	28	46.7
3:<6	16	26.7
6:<9	12	20
9: ≤ 12	4	6.7
The disease was discovered by		
Chance	11	18.3
Signs and symptoms of the disease	12	20
Investigations	30	50
Complications of the disease	7	11.7
Number of follow up per month		
Once	41	68.3
Twice	19	31.7

Table 2. Number and percentage distribution of the studied children according to their history of the disease (n=60).

Variables	Mother		Father	
	No.	%	No.	%
Age in years				
20<25	6	10	3	5
25<35	38	63.3	20	33.3
35 ≤ 45	16	26.7	37	61.7
X ± SD	32.90 ± 6.17	36.85 ± 6.59	30	30
Level of education				
Illiterate	6	10	6	10
Read and write	3	5	2	3.3
Basic education	9	15	7	11.7
Technical education	30	50	31	51.7
High education	12	20	14	23.3
Occupation				
Working	14	23.3	52	86.7
Not working	46	76.7	8	13.3

Table 3. Number and percentage distribution of the studied parents according to their characteristics (n=60).

Total level of children's knowledge	Pre	Post	5	5	X ²	p-value
	No.	%	No.	%		
Satisfactory	15	25	49	81.7	5.532	0.019*
Unsatisfactory	45	75	11	18.3		

Note: (*): Statistical significance difference, p<0.05.

Table 4. Number and percentage distribution of the studied children according to their total level of knowledge regarding phenylketonuria pre/post nursing intervention (n=60).

Total level of depression	Post		Post		X ²	p-value
	No.	%	No.	%		
Mild (1:<18)	20	33.3	47	78.3	15.894	0.000***
Moderate (18:<36)	35	58.3	13	21.7		
Severe (36: ≤ 54)	5	8.3	0	0		

Note: (***): Extremely statistical significance difference, p<0.0001.

Table 5. Number and percentage distribution of the studied children according to their total level of depression pre/post nursing intervention (n=60).

Total level of anxiety	Pre program		Post program		X ²	p-value
	No.	%	No.	%		
Mild (1:<12)	7	11.7	39	65	38.198	0.000***
Moderate (12:<24)	23	38.3	21	35		
Severe (24: ≤ 36)	30	50	0	0		

Note: (***): Extremely statistical significance difference, p<0.0001.

Table 6. Number and percentage distribution of the studied children according to their total level of anxiety pre/post nursing intervention (n=60).

Table 6 cleared that there was extremely statistical significant difference between the studied children's level of anxiety pre and post nursing intervention implementation, where half of the studied children (50%) had severe level of anxiety pre nursing intervention implementation, while post nursing intervention implementation more than two thirds of them (65%) had mild level of anxiety.

Table 7 represented that there was highly statistical significant difference between the studied children's level of loneliness pre and post nursing intervention implementation, where about two thirds of the studied children (61.7%) had moderate level of loneliness pre nursing intervention implementation, while post nursing intervention implementation most of them (80%) had mild level of loneliness.

Table 8 represented that more than two thirds of the studied children (66.7%) had low level of self-esteem pre nursing intervention implementation, while post nursing intervention implementation about three quarters of them (71.7%) had high level of self-esteem.

Figure 1 the minority of the studied children (11.7%) always adjusted to their home pre nursing intervention implementation. Meanwhile, this percentage improved to be about two thirds (61.7%) post nursing intervention implementation.

Figure 2 indicated that about one third of the studied children

(28.3%) always adjusted to their school pre nursing intervention implementation, while post nursing intervention implementation this percentage improved to more than three quarters of them (76.7%).

Figure 3 revealed that the minority of the studied children (11.7%) always adjusted socially pre nursing intervention implementation, while post nursing intervention implementation most of them (80%) always adjusted.

Figure 4 revealed that the minority of the studied children (16.7%) always adjusted toward compliance with their medication pre nursing intervention implementation, while post nursing intervention implementation about three quarters of them (70%) always adjusted.

In Table 9 and Figure 5 shows that, there was statistical significance difference regarding to total adjustment patterns of the studied children pre and post nursing intervention implementation. It was cleared that the minority of the studied children (16.7%) always adjusted pre nursing intervention implementation, while post nursing intervention implementation about three quarters of them (71.7%) always adjusted.

Table 10 showed that, there was statistical significant difference between the studied children's characteristics and their total level of adjustment patterns. It was observed that 43.2%, 21.7%, 37.6%, 28.3%, 35.0% and 25.0% of the studied children who

Total level of loneliness	Pre		Post		X ²	p-value
	No.	%	No.	%		
Mild (1<13)	5	8.3	48	80	14.645	0.001***
Moderate (13<26)	37	61.7	12	20		
Severe (26 ≤ 40)	18	30	0	0		

Note: (***): Extremely statistical significance difference, p<0.0001.

Table 7. Number and percentage distribution of the studied children according to their total level of loneliness pre/post nursing intervention (n=60).

Total level of self esteem	Pre		Post		X ²	p-value
	No.	%	No.	%		
Low (<13)	40	66.7	0	0	5.388	0.068
Moderate (13:<26)	14	23.3	17	28.3		
High (26: ≤ 40)	6	10	43	71.7		

Table 8. Number and percentage distribution of the studied children according to their total level of self-esteem pre/post nursing intervention (n=60).

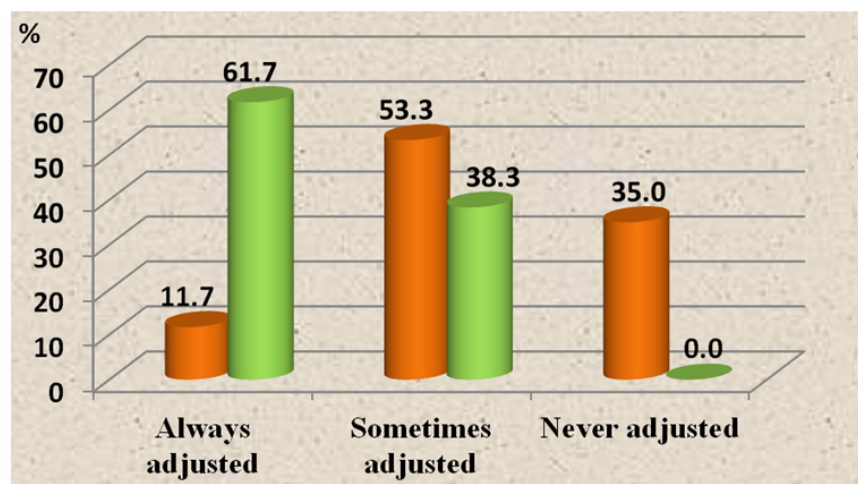


Figure 1. Percentage distribution of the studied children’s total home adjustment pre/post nursing intervention (n=60). Note: Pre-intervention: (orange); Post-intervention: (green).

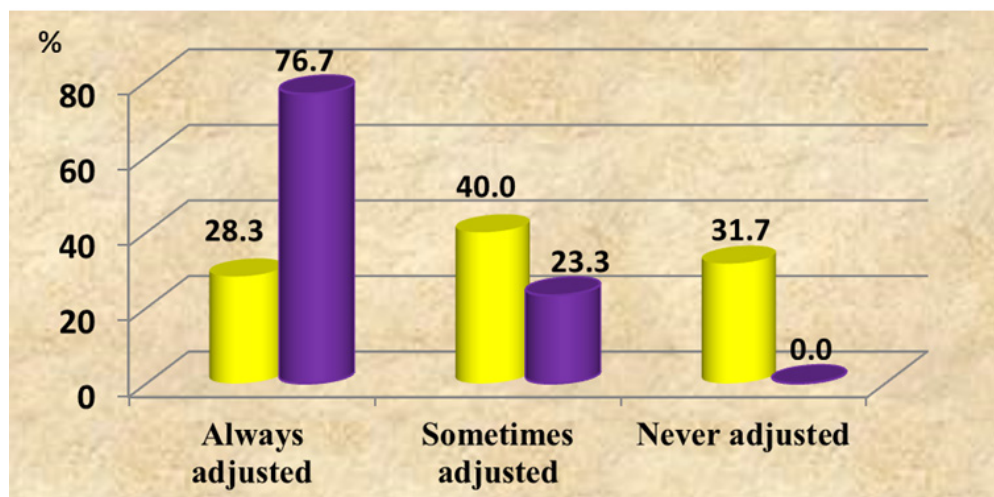


Figure 2. Percentage distribution of the studied children’s total school adjustment pre/post nursing intervention (n=60). Note: Pre-intervention: (yellow); Post-intervention: (purple).

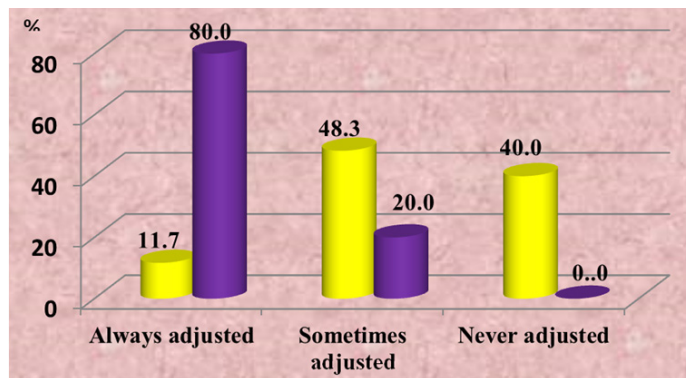


Figure 3. Percentage distribution of the studied children’s total social adjustment pre/post nursing intervention (n=60). Note: Pre-intervention: (■); Post-intervention: (■).

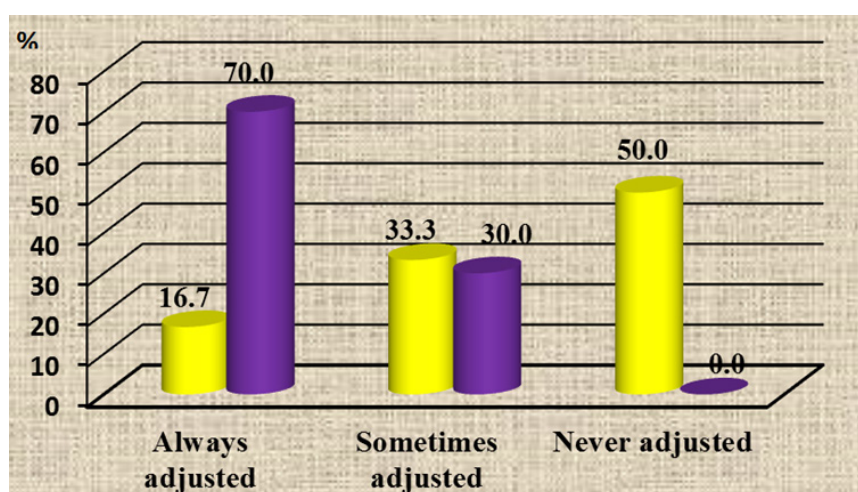


Figure 4. Percentage distribution of the studied children’s total compliance with their medication pre/post nursing intervention (n=60). Note: Pre-intervention: (■); Post-intervention: (■).

Total level of self esteem	Pre		Post		X ²	p-value
	No.	%	No.	%		
Always adjusted	10	16.7	43	71.7	34.708	0.003*
Sometimes adjusted	27	45	17	28.3		
Never adjusted	23	38.3	0	0		

Table 9. Number and percentage distribution of the studied children’s total adjustment patterns pre/post nursing intervention (n=60).

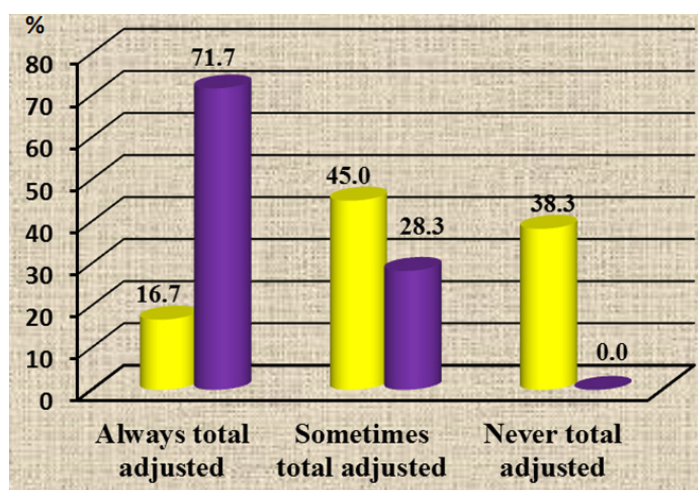


Figure 5. Percentage distribution of the studied children’s total adjustment patterns pre/post nursing intervention (n=60). Note: Pre-intervention: (■); Post-intervention: (■).

Total children's adjustment patterns																
Children's characteristics	Pre						Post									
	Never n=23		Sometimes n=27		Always n=10		X ²	p-value	Never n=0		Sometimes n=17		Always n=43		X ²	p-value
	No.	%	No.	%	No.	%			No.	%	No.	%	No.	%		
Age in years																
6:<8	4	6.7	1	1.7	0	0	26.29	0.000***	0	0	5	8.3	0	0	34.2	0.000***
8:<10	10	16.7	0	0	0	0			0	0	8	13.3	2	3.3		
10: ≤12	9	15	26	43	10	16.7			0	0	4	6.7	41	68.3		
Gender																
Male	13	21.7	13	22	7	11.7	0	0	0	0	11	18.3	22	36.7	14.9	0.003*
Female	10	16.7	14	23	3	5			0	0	6	10	21	35		
Level of education																
Not yet enrolled	1	1.7	1	1.7	0	0	14.49	0.006*	0	0	2	3.3	0	0	15.7	0.004*
Primary	19	31.6	19	32	10	16.7			0	0	14	23.3	34	56.7		
Preparatory	3	5	7	12	0	0			0	0	1	1.7	9	15		
Birth order																
First	15	25	17	28	7	11.7	11.41	0.016*	0	0	12	20	27	45	11.3	0.17*
Middle	8	13.3	9	15	1	1.7			0	0	5	8.3	13	21.7		
Last	0	0	1	1.7	2	3.3			0	0	0	0	3	5		
Residence																
Urban	7	11.7	6	10	1	1.7	11.66	0.02*	0	0	3	5	11	18.3	10.8	0.03*
Rural	16	26.6	21	35	9	15			0	0	14	23.4	32	53.3		
Child's age at diagnosis of the disease in years																
<3	9	15	15	25	4	6.7	13.322	0.009*	0	0	5	8.3	23	38.3	13.25	0.01*
3:<6	6	10	6	10	4	6.7			0	0	6	10	10	16.7		
6:<9	6	10	4	6.7	2	3.3			0	0	0	0	0	0		
9: ≤12	2	3.3	2	3.3	0	0	0	0	0	0	0	0	0	0	0	0
The disease was discovered by																
Chance	5	8.3	4	6.7	2	3.3	14.241	0.01*	0	0	4	6.7	7	11.7	12.45	0.015*
Signs and symptoms	7	11.7	4	6.7	1	1.7			0	0	5	8.3	7	11.7		
Investigations	9	15	16	26.7	5	8.3			0	0	7	11.7	23	38.3		
Complications	2	3.3	3	5	2	3.3	0	0	1	1.7	6	10	0	0	0	0
Number of follow up/month																
Once	16	26.7	16	26.7	9	15	13.213	0.014*	0	0	11	18.3	30	50	10.94	0.02*
Twice	7	11.6	11	18.3	1	1.7			0	0	6	10	13	21.7		

Note: (***) : Extremely statistical significance difference, p<0.0001. (*): Statistical significance difference, p<0.05.

Table 10. Relation between the studied children's characteristics and their total level of adjustment patterns pre/post nursing intervention (n=60).

were in the age group of 10: ≤ 12 years, males, in the primary stage, first child, came from rural residence and diagnosed with phenylketonuria in age less than 3 years were sometimes able to adjust pre nursing intervention, while 68.3%, 36.7%, 56.7%, 45.0%, 53.3% and 38.3% of them were always able to adjust post nursing intervention with statistical significant difference ($X^2=34.172, 14.903, 15.731, 11.303, 10.752$ and 13.248 at $p<0.05$).

Discussion

In relation to the characteristics of the studied children, it was observed from Table 1 that, three quarters of the studied children were aged 10: ≤ 12 years ($X \pm SD 10.16 \pm 1.51$ years). This study finding contradicted with a study conducted in Egypt by Ibraheem et al. [7] entitled "Mother's coping of children suffering from phenylketonuria" reported that, more than half of the studied children were aged 2<6 years.

As regards to the gender of the studied children, the results of the current study showed that, more than half of the studied children were males. This result was similar to the results of study by Fouad et al. [8] which entitled "Nursing intervention program for family caregivers having children with phenylketonuria" who found that, more than half of the studied children (55.6%) were males. On the other hand, this study finding contradicted with a study conducted in Australia by Morawska et al. [10] entitled "Psychosocial functioning in children with phenylketonuria: relationships between quality of life and parenting indicators" reported that, more than two thirds of the studied children (66.7%) were females.

The present study revealed that, most of the studied children were in primary school and almost two thirds of them were ranked as the first order. This result was in an agreement with Elsayed et al. [11] study which entitled "Assessment of mothers care toward their children having Phenylketonuria" who found that, about half of the studied children (43.6%) were in primary school and more than one third of them (42.6%) were ranked as the first child in their families.

The result of the current study revealed that, more than three quarters of the studied children came from rural areas. This result was consistent with Ahmed et al. [3] study that entitled "Pediatric phenylketonuria in Fayoum governorate retrospective study" who found that, the majority of the studied children (95.9%) were from rural area. From the researcher point of view, this could be due to lack of health settings in rural areas.

In relation to the history of children' disease, it was noticed from Table 2, that about half of the studied children were diagnosed with phenylketonuria in in the age less than 3 years and half of them discovered the disease by investigations [12]. Regarding the number of follow/month, it was found that more than two thirds of the studied children had follow up visits once/month. This result was supported with Fouad et al. [8] who found that the age at disease detection was from 1<3 years for 45.7% of the studied children. Regarding discovery of the disease, most of them (88.2%) discovered the disease from investigation and for 79.7% of them the follow up frequency was once per month.

According to characteristics of the studied children's parents, it was observed from Table 3 that, half of the studied parents had technical education [9]. The study finding was similar with Elsayed et al. [11] who revealed that about half of the studied parents had intermediate education. Concerning the occupation of the studied mothers, the findings showed that, more than three quarters of them were not working, this result come in an agreement with Ibraheem et al. [7] who found that, the majority of the studied mothers of the children having phenylketonuria (92%) were not working. Also Etemad et al. [12] in a study entitled "Health-related quality of life of parents of children with phenylketonuria in Tehran province, Islamic republic of Iran", who found that, about half of the studied sample (49.8%) were housewives.

Regarding to occupation of the studied fathers, the findings showed that, the majority of the studied fathers were working. This study finding is congruent with what reported by Morawska et al. [10] who found that 83.3% of the studied fathers were working while only 16.7% of them were not working. The researcher believed that this finding may be due to fathers in Egypt are the main source for monthly income in their families so the majority of fathers are working.

There was statistical significant difference between the studied children's total level of knowledge pre and post nursing intervention implementation Table 4. It was noticed that three quarters of the studied children had unsatisfactory knowledge regarding phenylketonuria pre nursing intervention as compared with most of them had satisfactory knowledge post nursing intervention. This result was supported with Fouad et al. [8] who found that there were highly statistically significant differences between the studied sample's knowledge post-test compared to pre-test regarding the meaning of PKU, the causes of this disease, its signs and symptoms, the different methods of treatment, prevention of potential complications of the disease. From the researcher point of view, this result could be due to the fact that school age children had intellectual curiosity to know everything about their disease and its medication, which they acquainted enough about it through the nursing intervention program.

Regarding to the total level of depression of the studied children, it was cleared from Table 5 that, there was extremely statistical significant difference between the studied children's total level of depression pre and post nursing intervention implementation, this result was supported with Ford et al. [13] who found that rates of depression are significantly higher among children with PKU than the general population. Depression is reported by more than half of the studied sample in survey data. From the researcher point of view, this result could be due to the phenylketonuric children perceived themselves as being different and their inability to communicate with others.

Regarding to the total level of anxiety of the studied children, it was noticed from Table 6 that, half of the studied children had severe anxiety pre nursing intervention implementation. From the researcher point of view, this might be due to lack of health education about phenylketonuria which led to those children that they are handicapped and not able to live as their peers. This result came in agreement with Ashe et al. [14] in a study entitled "Psychiatric and cognitive aspects of phenylketonuria: The

limitations of diet and promise of new treatments", who reported that; rates of anxiety disorders are also significantly higher in the overall PKU population compared to the general population.

However, in the current study after nursing intervention implementation, a significant improvement in the studied children's level of anxiety was obvious. From the researcher point of view, this result could be due to continuous education of phenylketonuric children by providing information focusing on the disease and its associated problems as anxiety, is essential for positive improvement of anxiety level.

In relation to the total level of loneliness of the studied children, it was cleared from Table 7 that, about three quarters of the studied children always feel strange, isolated from others and social relationships are superficial pre nursing intervention implementation. These findings are similar to a study conducted by Jahja et al. [15] which entitled "Social-cognitive functioning and social skills in patients with early treated phenylketonuria: A PKU-COBESO study", who found that, PKU-patients showed poorer social-cognitive functioning and reportedly had poorer social skills. From the researcher point of view, this result might be due to the fact that phenylketonuric children might have been exposed to peers curiosity about their illness, which led the child had feelings of being different from others.

Regarding to the total level of self-esteem of the studied children, it was cleared from Table 8 that, more than two thirds of the studied children had low self-esteem pre nursing intervention implementation, while post nursing intervention implementation about three quarters of them had high self-esteem. This result came in agreement with Iakovou Schulpis et al. [16] in a study entitled "Self-estimation of phenylketonuria patients on therapeutic diet, psychological support" who reported that; more than two thirds of the studied children had moderate and low self-esteem before psychological support. From the researcher point of view, this result might be due to the fact that phenylketonuric children develop a sense of inferiority and perceive themselves as being different before nursing intervention implementation.

Regarding to total home adjustment of the studied children, it was noticed from Figure 1 that, the minority of the studied children always adjusted to their home pre nursing intervention implementation. Meanwhile, this percentage improved to be about two thirds post nursing intervention implementation. From the researcher point of view, this result could be due to three quarters of the studied children were aged $10 \leq 12$ years, which enable them to understand more about their illness and realize the importance of carrying out medical instructions and parents' advices [17].

As regards to total school adjustment of the studied children, It was noticed Figure 2 that only one third of the studied children able adjust to their school pre nursing intervention implementation, while post nursing intervention implementation this percentage improved to more than three quarters of them. This result came in agreement with Ashe et al. [13] who found that, children with PKU commonly demonstrate school difficulties, they face many challenging situations, demanding of executive skills and goal-directed behavior. Therefore, children with PKU may require additional educational support and counseling to help compensate for executive difficulties. From the researcher point

of view, this result could be due to the phenylketonuric children feel different than others as a consequence of being ill. So, this feeling may prevent them from interaction with their peers and teachers. However post nursing intervention, they viewed it differently [18].

In relation to total social adjustment of the studied children, it was cleared from Figure 3 that, minority of the studied children always adjusted socially pre nursing intervention implementation, while post nursing intervention implementation most of them always socially adjusted. This result was supported with Ford et al. [12] who found that most of children with PKU withdrew from social situations and became socially isolated [19].

As regard to total adjustment patterns of the studied children in the form of compliance with their medication Figure 4 showed that three quarters of the studied children obeyed medical order regarding to illness and treatment plan and were optimistic and feeling that medication will cause recovery post nursing intervention implementation [13]. From the researcher point of view, this result could be due to awareness of phenylketonuric children about how the benefits of medical compliance, in addition to children in this age are learning to master independence; they are creative, sociable and develop sense of competence [20].

Concerning to the relation between the studied children's total level of adjustment patterns and their characteristics, the current study in Table 10 showed that, there was statistical significant difference between the studied children's characteristics and their total level of adjustment patterns, where the studied children who were in the age group of $10 \leq 12$ years, males, in the primary stage, first child, came from rural residence were sometimes able to adjust [21].

Conclusion

The study concluded that, there was a positive effect of the nursing intervention on the adjustment patterns of children suffering from phenylketonuria. Also, there was an extremely statistical significant difference and positive correlation between children's total knowledge and total adjustment patterns pre and post nursing intervention implementation. There was there was a positive effect of the nursing intervention on the adjustment patterns of children suffering from phenylketonuria. The study findings revealed that there was statistical significant difference between the studied children's knowledge pre and post nursing intervention implementation.

Recommendations

In the light of the study findings, the following recommendations are suggested

- Continuous health educational programs about adjustment patterns should be provided for children suffering from phenylketonuria to cope effectively with their disease.
- Emphasizing the importance of continuous assessment of children suffering from phenylketonuria for early detection of complications and proper management.
- Designing an educational handout for children about phenylketonuria, its management and prevention of its related

complications.

• Further researches are required involving larger study sample of children suffering from phenylketonuria to improve their knowledge, attitudes, and practices and empowering their adjustment patterns positively.

Ethical Considerations

Verbal approval from parents and their phenylketonuric children was a pre-requisite to include the child in the study sample. They were informed that all the gathered data were used for the research purpose only. The study subjects were informed about the purpose and expected outcomes of the study and they were assured that the study is harmless and their participation is voluntary. They had the right to withdraw from the study at any time. They were assured also that anonymity and confidentiality were guaranteed.

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***Correspondence to**

Manar Fathi Hassan
Department of Pediatric Nursing
Faculty of Nursing
Helwan University, Cairo
Egypt
E-mail: sara.galal@nursing.helwan.edu.eg