The Effects of Coping Strategies Intervention on Strain and Quality of Life among Mothers of Children with Phenylketonuria

[1]Fatma A. Eiz - Elregal , [2] Aml Reda Mohamed Ebrahem Mansy [3] Enaam Abd-El latif Farrag Hamza [4] Mayada Taha Mahmoud Sabea

[1] Assistant Prof of Community Health Nursing, Faculty of Nursing (for girls)-Al-Azhar University, Egypt.
[2] Lecturer of Pediatric Nursing, Faculty of Nursing- Mansoura University, Egypt.
[3] Assistant Prof of Psychiatric Mental Health Nursing, Faculty of Nursing, - Fayoum University, Egypt.
[4] Assistant Prof of Community Health Nursing, Faculty of Nursing, Helwan University, Egypt.

Abstract

Background: Phenylketonuria is often called PKU. Mothers face multiple problems because they are playing a vital role in caring for sick children for long time, thus helping decreasing strain and promoting health for maintain their wellbeing. **Aim**: This study aimed to evaluate the effects of coping strategies intervention on strain and quality of life among mothers of children with phenylketonuria. **Method:** a quasi-experimental design was conducted in out-patient clinics and genetics department affiliated to Mansoura University Children's Hospital (MUCH), involving 48 mothers of children diagnosed with phenylketonuria. Data were collected using a structured interviewing questionnaire sheet, mother knowledge about PKU, Coping Inventory for Stressful Situation (CISS), Modified Caregiver Strain Index (MCSI) and the WHO Quality of Life-BREF (WHOQOL-BREF). **Results:** The current study reveals that enhancing coping strategies after applying the study intervention. The mean of mothers 'coping strategies in posttest was higher than in the pretest (193.43 ± 9.26 & 79.8±7.2respectively) with a statistically significant difference between the two-study intervention. Also, there was a highly statistically significant negative correlation between pre- and post- coping score and strain level ($r=-0.298$, $p<0/001$; $r=-0.401$, $p<0.001$) and between pre and post quality of life and strain level among mothers having children with PKU ($r=-0.234$, $p<0.001$; $r=-0.352$, $p<0.001$) and the positive correlation between coping and QOL pre and post intervention ($r=0.332$, $p<0.001$; $r=0.463$, $p<0.001$). **Conclusion:** The study concluded that, coping strategies intervention had a positive effect on reducing strain and improving quality of life among mothers of children with PKU. **Recommendations:** the specialist can educate the mother how to adapt by using coping strategies when caring child with PKU, especially those who have a positive family history and doing regular training about using coping strategies.

Keywords: Mothers, Children with Phenylketonuria, Quality of Life, strain, Coping Strategies.

Introduction

Phenylketonuria (PKU) is a rare autosomal recessive disorder which causes toxic buildup of phenylalanine, cause irreversible brain damage if untreated and leading to microcephaly, seizures, severe intellectual disability and behavioral problems in untreated individuals. **(Lingtao & Zhang, 2023)**. Phenylketonuria (PKU) is an inborn error of metabolism (IEM) most often caused by missense mutations in the gene encoding phenylalanine hydroxylase (PAH), which catalyzes the hydroxylation of phenylalanine (Phe) to generate tyrosine **(Hillert et al ., 2020; Mojibi, Ghazanfari-Sarabi & Hashemi-Soteh, 2021)**.

It is chronic disease affected newborn and may lead to brain damage or even death unless therapy is initiated within two weeks of birth. PKU cannot be cured, but can be effectively treated if a special diet is started early in the newborn period. Current researches indicate that the special diet should be continue as eliminating high-protein foods like eggs, milk, cheese, nuts, and fish, and control their intake of bread, rice, pasta, and some vegetables through adolescence and possibly throughout life **(MacDonald et al., 2020).** Recent studies discovered that PKU occurs in a child who has two genes for PKU, one inherited from their father and the other from their mother **(Van Spronsen et al., 2021)**.

Studies conducted on mothers who communicating with, and educating the child with chronic diseases example PKU have shown that they bear many burdens such as household responsibilities, taking care of the rest of the healthy children, and taking care of the husband, and this increases the physical, psychological, mental and moral health burdens, in addition to self-neglect. **(Kargar, & Asgari Ebrahimabad, 2015; Ford, O'Driscoll & MacDonald, 2018)**.

Families' strain is the perception of persistent problems and a feeling of decreased wellbeing that results from providing prolonged care, often the mother provided continuous care for children with PKU, that lead to irreversible harm. Having a child with phenylketonuria is detrimental to the mental health of the family especially mother that may experience a higher level of strain, anxiety, and higher risk of fatigue in comparison with parents of normal children **(Ghahremani, Doulabi, Eslami, & Shekarriz-Foumani, 2107)**.

There are many situations that can cause strain on the caregivers' role. The financial burden of caring for a loved one can cause enormous strain, as the sudden increase in responsibility. Many people, especially younger mothers, can feel overwhelmed due to the sudden change in roles and a drastic shift in family dynamics **(Davidson, 2022)**. Quality of life is a health indicator, a holistic approach and a broad-ranging concept affected in a complex way by the individual's physical health, emotional state, social interaction, level of independence, and their relationships to environment **(Vanz, Félix, da Rocha & Schwartz, 2015)**. Various studies have focused on investigation of the quality of life of mothers of children with chronic diseases, given that the mother is the primary caregiver for chronic diseases that are considered incurable. As the child's illness progresses, this may affect the quality of life of caregivers' physical, emotional, social, and financial well-being. **(Sheng, Ma, Ding & Zhang, 2019)**.

Coping strategy intervention deal with and attempt to overcome problems and difficulties. These may help a mother face a situation, take action, and be flexible, and manage crises. It consists of three types, the first one is practical methods of the task oriented strategy as problem focused coping strategies directly address the problem in an effort to eliminate the stressor and collecting more information to solve the problem, consultation and attention to the positive points, the second one is emotional-focused coping refers to efforts that reduce the degree of emotional distress induced by the stressful situation and consists of obsession, fantasy, anger, crying, feeling loneliness, depression, and the third the avoidant strategy involves avoiding the issue and problem and, in some cases, seeking social support or turning to social entertainment **(Choi et al., 2017; Kenneson, Youngborg & Singh, 2020)**.

Mothers play a pivotal role in managing PKU in childhood resulting in a lack of time for self-care, financial stress, and feeling overwhelmed or unsupported in parenting roles. Symptoms may include fatigue, difficulty concentrating, irritability, stress and other difficult situations. Therefore, mothers should receive support from nurses and other health care professionals all the time to enable them to cope with care problems and allow them to better cope with pressures to adopt the necessary skills, improve the quality of life and develop new behavior by adopting coping interventions **(Deek et al ., 2016)**.

Nurses are proactive in reducing stress and anxiety, providing emotional support during difficult times for mothers by understanding, listening to reduce stress, relaxing, providing practical assistance, creating solutions to make life easier and developing coping strategies that are tailored to meet their need. Also, children suffering from long-term conditions require continuing support and nursing care throughout their lives (**Khalil Ibrahim, Amin Mohamed, Salah Eldin AlRafay & Ali Khalifa, 2023; Sheth et al., 2023)**.

Significance of the study

PKU has an estimated prevalence of 1 in 23,930 live births worldwide and affects about 0.45 million individuals, of whom at least two-thirds require treatment while in Egypt, every year affected with PKU as one million babies

are born yearly **(Araby, Fateen, & Gouda, 2009; Hillert et al., 2020)**. Higher rates in Lower Egypt and South Sinai are due to increase the rate of consanguinity to 50% and PKU systematic neonatal screening program is lacking when women with PKU have high blood phenylalanine levels during pregnancy, it can harm their unborn baby. Infants and young children are the future of the country, the healthy growth and development of them is the paramount importance for them to develop full physically and mental potentials development **(El-Metwally et al., 2018)**.

Aim of the Study

This study aimed to evaluate the effects of coping strategies intervention on strain and quality of life among mothers of children with phenylketonuria through:

1.Assess mothers' knowledge about PKU
2.Assess caregivers' strain level of mothers of children with phenylketonuria
3.Assess quality of life of mothers of children with phenylketonuria.
4.Assess coping strategies of mothers of children with phenylketonuria.
5.Implement the coping strategies intervention based on an assessment of mother's knowledge, strain and quality of life.
6.Evaluate the effect of the coping strategies intervention on mother's knowledge, strain and quality of life.

Research Hypothesis:

Applying the coping strategies intervention for mothers of children with phenylketonuria will decrease strain level and improve knowledge and quality of life after intervention than before.

Method

Research design:

A quasi-experimental research design aims to establish a cause-and-effect relationship between an independent (coping strategies) and dependent variable (knowledge, strain level and quality of life) (one group pre/posttest) was utilized to achieve the aim of the study.

Settings:

The current study was carried out at out-patient clinics and genetics department at Mansoura University Children's Hospital (MUCH) in Mansoura City, Dakahlia Governorate, Egypt

Sampling:

A purposive sample of 48 mothers of children with a confirmed diagnosis of phenylketonuria was used in this study. During 4 months of participation in the study the inclusion in the study sample whenever they met the following criteria.

Inclusion criteria:

Had Children's age ranged from 4 to less than 12 years from both genders.

Exclusion criteria:

Had Children suffering from any other disorders such as (ADHD, mental retardation, epilepsy, psychosis, etc.……)

Tools of data collection:

Four tools were used in this study to collect the data, the first tool developed by the researchers after reviewing the national and international related literature.

The tool I: A structured interview questionnaire (pre/posttest) was developed in a simple clear Arabic language by the researchers based on a literature review **(Chandler & Venditti, 2016)** and experts' opinions in light of relevant references to assess mothers' knowledge regarding phenylketonuria. It consisted of two parts. **It was included data about:**

Part 1: Demographic characteristics of mothers of children with phenylketonuria such as age of mother, level of education, occupation, economic status and number of children and also include characteristics of their children such as; gender age, residence, and family history of phenylketonuria.

Part 2: Mother's knowledge (pre-posttest format) regarding applying phenylketonuria caregiver care which consisted of 14 open end questions on PKU disease and diet.

Scoring system fourteen questions were used to test mother's knowledge about PKU, with a total of 28 points, a correct complete answer was scored (2) and a correct incomplete answer was scored (1), while the wrong answer was given (0), according to the mothers' answers, her knowledge score was categorized into (Good knowledge) ≥ 75%, (Average knowledge) ≥ 50% - < 75% and (Poor knowledge) < 50%.

Tool II: Coping Inventory for Stressful Situation (CISS): The scale was adapted from **Park, Kim, & Noh, 2000**. CISS is a 48-item self-report inventory that measures stress coping strategies in three dimensions-namely task oriented coping (16 items), emotion-oriented (16 items), and avoidant strategy (16 items). Task oriented coping refers to activities through which problems are directly confronted and solved; Practical methods of the task-oriented strategy include using past experience, realistic, collect more information to solve the problem, consultation and attention to the positive points. Emotional-focused coping refers to addressing emotional responses and efforts that reduce the degree of emotional distress induced by the stressful situation and consists of obsession, fantasy, anger, crying, feeling loneliness, depression and other conscious activities. The avoidant strategy involves avoiding the issue and problem and, in some cases, seeking social support or turning to social entertainment.

Scoring system: each item is based on a 5-point Likert scale (always = 5, often= 4, usually= 3, sometimes= 2, never= 1). The higher score in each of these three subscales categories determine the dominant strategy of every individual. The lowest and highest scores in the task-oriented coping were 16 and 80 respectively; the lowest and highest scores in the emotion-oriented coping were 16 and 80, respectively. The lowest and highest scores in the in the avoidant strategy, were 16 and 80. The dominant strategy of each mother is determined by his/her score in each of the three dimensions of coping strategies, that the mother's coping strategies was considered adequate: if the percent score was ≥ 60%, while it was considered inadequate, if the percent score was less than 60%.

Tool III: the Modified Caregiver Strain Index (MCSI): The scale was adapted from **Thornton & Travis, 2003**. It contained13 items to assess caregiver strain related to care. There is at least one item for each of the following major domains: Employment, Financial, Physical, Social, and Time. Scoring system: Sum responses for "Yes, on a regular basis" (2 pts each), "yes, sometimes" (1 pt each) and "no" (zero pt each) then summing across all scale items. Total score ranged from 13- 26

►Scores ranging from 0 -> 13 would be considered low strain.

►score ranging from 14->20 would be considered moderate strain

►Scores ranging from ≤ 20 would be considered high level of strain.

Tool IV: the WHOQOL-BREF is a 24-item, self-administered, generic questionnaire that is a short version of the WHOQOL-100 scale was developed by **World Health Organization, 1993,** to assesses an individuals' perceptions of their quality of life in the context of culture, value systems, goals and concerns divided into 4 categories; the Physical Health (7 items), Psychological Health (6 items), Environmental Health domain (8 items), and social relationships (3 items). Individual scores can be found for each category. Every question in the domain was given on a five-point Likert scale, with the following possible answers: (1) minimal, (2) little, (3) moderate, (4) very much, and (5) extreme.

Scoring system: subsequently, the mean score for each domain is calculated, resulting in a mean score per domains that is between 5 and 25. Finally, this mean domain score is then multiplied by 4 in order to transform the domain score into a scaled score. A method for manually calculating individual scores for selected questions from the WHOQOL-100 (abbreviated to WHOQOL-BREF24). is below:

Physical domain= ((6-Q3) + (6-Q4) + Q10 + Q15 + Q16 + Q17 + Q18)) x4.

Psychological domain= (Q5 + Q6 + Q7 + Q11 + Q19 + (6-Q26) x4.

Social Relationships domain= (Q20 + Q21 + Q 22) x4.

Environment domain= (Q8 + Q9 + Q12 + Q13 + Q14 + Q23 + Q24 + Q 25) x4, with a higher score indicating a higher QOL. When transformed by multiplying x4, each domain score is then comparable with the scores used in the original WHOQOL-100.Mothers score was classified as follows: poor quality of life score was less than 50% represent >50 marks, average quality of life score of mothers equal 50 marks, good quality of life mothers scores more than 50% present<50 marks. The Arabic translation of WHOQOL-BREF was by **Ohaeri &Awadalla ,2009**. Using forward-back translation.

Validity and Reliability

The tools were evaluated for appropriateness, completeness, and legibility by an expert panel consisting of two professors from the community health nursing department, one professor from the psychiatric mental health nursing department and two professors from pediatric health nursing - Faculty of Nursing. The panel ascertains the face and content validity of the tools. The reliability was done by using Cornbrash's Alpha coefficient test which revealed that each of the three tools consisted of relatively homogenous items as indicated by the moderate to the high reliability of each tool. The internal consistency of knowledge was (0.85), the total 13-items care giver strain scale was (0.91). The total quality of life was (0.89) and the internal validity of the coping strategies obtained at high levels (0.86) using Cronbach's alpha. Pearson's correlation coefficient obtained at 0.85 for task-oriented strategy, 0.83 for emotion-oriented, 0.81 for avoidant.

Ethical consideration

Official permissions to conduct the proposed study were obtained from the Scientific Research Ethical Committee, Faculty of Nursing, El-Mansoura University (Ref.No. P 0630). All mothers were informed about the importance and aim of this study. All mothers were informed that their participation is voluntary and their rights to withdraw at any time, and confidentiality of the information obtained. Also, the mothers were informed that the collected data would be used only for the purpose of the present study, as well as for their benefit.

Pilot study

It will be done on 10% mothers of children with PKU (5) mothers of children with PKU, to evaluate the feasibility, applicability, and clarity of the research tools and to estimate the time needed for data collection, and the feasibility of the research process. Based on results, the modification done. The subjects were excluded from the study to ensure validity of final results.

Procedure

The study lasted about 4 months from the beginning of September 2024 to the end of December 2024. - The researchers attended out-patient clinics and genetics department two days per week (Monday and Thursday) about three weeks from 9 a.m. to 12 p.m. to collect data and assess the needs (pre-test). The contents of the intervention program were designed according to literature **(Park, Kim & Noh, 2000)**. The researchers chose the mothers based on the inclusive criteria of the study.

Assessment phase (pretest)

The researchers interviewed each mother individually to obtain written consent after providing explanations about the purpose and nature of the study, and to ensure data confidentiality. In the pre-test phase, the researchers stayed with each mother and allowed her to answer all questions according to four tool sheets. This phase aimed to assess mothers' knowledge about PKU, coping strategies, strain, and quality of life. Based on the assessment results the researchers applied the coping strategies intervention from the beginning of September to the end of December 2024.

1.Planning phase

The researchers developed a simple, colorful Arabic strategies intervention for, drawing on relevant literature and the findings from the pre-test. The intervention was

specifically tailored to address the needs of the mothers of children with PKU. The researchers organized the intervention into fifth educational sessions

2.Implementation phase

The developed coping strategies intervention was implemented for the study group in the out-patient clinics and genetics department. It was conducted in five sessions for each group, the group consisted from eight mother. Each session took about 30 to 45minutes. Sessions were given to each mother in the study group to attract her attention, motivate her, and help for reviewing at home which highlighted the major points discussed in the sessions and detailed home practice assignments by using effective media of conveying information as, laptop, power point presentation and some videos to adapt with stressful situations as follows;

- **The first session** was providing knowledge about PKU definition, signs and symptoms, treatments, genetic factor, and diet).
- **the second session** was carried out in the same week for educating and training the mothers about task-oriented coping: Primary positive aspects of life that are adaptable when situations are assessed as modifiable, the goal is to solve the problem by training mothers on the essential points of task-oriented coping.
- **The third session** was carried out in the second week for educating and training the mothers about emotion-oriented coping: secondary control is applied when situations are assessed as challenging to change. The goal is to reduce stress and avoid negative outcomes such as strain and anxiety. Mothers were trained on how to apply the elements of emotional coping practice.
- **The fourth session** was carried out in the same second week for educating and training the mothers about avoidant coping: the goal is to focus on something more interesting and adaptive for short-term, but over time it is more effective to develop the skills needed to address the threat as an instrumental problem. Applying coping strategies to improve physical, emotional and mental health.
- **The fifth session** was carried out in the first third week for practicing coping strategies at the beginning of every session feedback about the previous session was done.

3.Evaluation phase

After one months from implementation of the coping strategies, each mother in the study group was interviewed to evaluate; her knowledge using tool I, evaluate her coping strategies using tool II and Caregiver strain and quality of life by using Tool III and tool IV

Statistical analysis

Statistical presentation and analysis of the present study was conducted, using the mean, standard deviation, chi-square test was used to compare between groups in qualitative data, paired Student T-test was used to compare between two means of the related sample and linear correlation coefficient was used for detection of correlation between two quantitative variables in one group by IBM SPSS Statistics for Windows, Version 28.0. Armonk, NY: IBM Corp.

Significant level

>0.05 Non-significant <0.05* significant <0.001* High significant

Results:

Table (1) shows demographic characteristics of studied subjects. This table shows that the highest percentage of mothers' age ≥ 30 years (52.1%) with a mean of 31.04±5.41years, 6.3% of them were illiterate, and 20.8 % of them had higher education, 60.4 % of them were working and 62.5 % had insufficient income. Regarding the number of siblings 35.4 % of them had one sibling, 33.3% of children were males and the children's age was ranged from 4 to less than 12 years with a mean of 8.54±2.46 years. Additionally, 50.0 % of them lived in rural areas and 58.3% from children had positive family history.

Table (2): Reveals the distribution of studied mothers` knowledge about

phenylketonuria disease and nutrition pre/posttest. Results indicate statistically significant improvement in mothers' knowledge regarding post intervention compared to pre intervention (Mean = 12.65±2.51& 24.09±2.75 respectively with t – test = 21.287, p< 0.05).

Figure (1): clarified that (54.2%) of the studied mothers had good total reported knowledge about PKU and nutrition in pretest, but posttest after coping strategies intervention, there was an improvement, (87.5 %) of them had a good total reported knowledge.

Table (3): shows that more than three quarter (79.2%) of mothers reported improvement in the coping strategies. In posttest were found improvement in the task-oriented coping, emotional coping and avoidant strategy (83.3%,77.1%, and 75.0% respectively) adequately compared by pretest (27.1%, 33.3%, and 29.2% respectively) insufficiently. With highly statistically significant differences between all coping strategies intervention among the studied mother's reported at pre- and post-intervention (p<0.001). Whereas mean of mothers 'coping strategies in posttest was higher than pretest (193.43 ± 9.26 versus 79.8±7.2 respectively).

Figure (2): showed that (29.2%) of the studied mothers had adequate coping strategies in pretest, but posttest, there was an improvement, in which (79.2% %) of them had adequate coping strategies.

Table (4) Clarifies that 25.0 % mothers had high strain in posttest compared to pretest. 72.9 % of mothers had high strain. There was a highly statistically significant difference between mothers' strain scores pre – post coping Strategies Intervention (X2=22.05 at P< 0.001). Whereas mean of mothers 'strain in posttest was lower than pretest (4.23±13 & 18.45±2.65 respectively).

Table (5) Shows that two third (64.6%) of mothers reported good total quality of life and in all dimension, physical, psychological, environment health and social relation (62.5%, 68.8%, 70.8% and 66.7% respectively), compared to pretest three quarter (75%) mothers reported poor total quality of life in all dimension, physical, psychological, environment health and social relation (87.5 %, 72.9%, 68.8% and 77.1%, respectively). There was a highly statistically significant difference between mothers' quality of life pre – post coping Strategies Intervention (X2=38.865 at P< 0.001) whereas mean of mothers 'quality of life in posttest was higher than pretest (86.15±5.84& 39.74±3.4 respectively).

Table (6): Shows that there was a significant positive correlation between total knowledge and total coping scores pre (r=0.427, p<0.001) and posttest (r= 0.372, p<0.001). Task-oriented coping, there was moderate positive correlation with total knowledge both pre (r= 0.430, p<0.001) and posttest (r=0.375, p<0.002). On the other, hand, Emotional coping also shows a significant positive correlation with total knowledge, both pre (r=0.384, p<0.001) and posttest (r= 0.425, p<0.001). Finally, avoidance coping demonstrates weaker but still statistically significant correlations both pre- (r = 0.198, p = 0.035) and posttest (r = 0.169, p = 0.015).

Table (7): Shows that there was a significant negative correlation between strain and coping scores pre (r=-0.298, p<0.001) and posttest (r=-0.401, p<0.001) and (strain and QOL scores) pre (r = -0.234, p=0.003) and posttest (r=-0.352, p<0.001). Furthermore, positively correlated with QOL and coping strategies intervention (r = 0.332, P < 0.001) in pretest and post-intervention r = 0.463, P < 0.001),

Table (8): shows that significant differences both pre- and posttest-intervention. Age significantly influenced coping abilities, with mothers under 30 showing a highly inadequate coping pre and posttest intervention (X^2 = 7.561, p = 0.006 & X^2 =4.703, p = 0.007). Mothers who could not read and write had insufficient coping skills before and after the intervention (X^2 = 9.379, p = 0.025& X^2 = 12.925, p = 0.005). On the other hand, the working mothers showing significantly positive coping abilities pre and post intervention (X^2 = 4.509, p = 0.105 & X^2 = 10.584, p = 0.005*).

Table (1): Distribution of socio-demographic characteristics of the studied subjects (n=48)

Variables	N	%
Age		
27 <30	23	47.9
≥ 30	25	52.1
Mean±SD	31.04±5.41	
Level of education		
Illiterate	3	6.3
Basic education	19	39.6
Intermediate education	16	33.3
Higher education	10	20.8
Job		
Housewife	19	39.6
Working	29	60.4
Income		
Sufficient	18	37.5
Insufficient	30	62.5
Number of Siblings		
1	17	35.4
2	14	29.2
3	12	25.0
4	5	10.4
Gender of child		
Male	16	33.3
Female	32	66.7
Age of child		
4-7 years	16	33.3
8 – less than 12 years	32	66.7
Mean±SD	8.54±2.46	
Residence		
Rural	24	50.0
Urban	24	50.0
Family history		
Yes	28	58.3
No	20	41.7

Table (2): Distribution of Mothers' knowledge about phenylketonuria disease and nutrition pre- and posttest coping Strategies Intervention (n=48).

	Pre						Post						Chi-square	
	Correct& complete		Correct& incomplete		Incorrect		Correct& complete		Correct& incomplete		Incorrect			
	N	%	N	%	N	%	N	%	N	%	N	%	t/X^2	P-value
What is the cause of phenylketonuria?	22	45.8	0	0.0	26	54.2	42	87.5	0	0.0	6	12.5	18.750	<0.001*
When is phenylalanine analyzed?	32	66.7	0	0.0	16	33.3	45	93.8	0	0.0	3	6.3	11.090	<0.001*
Is the missing enzyme in PKU phenylalanine hydroxylase?	19	39.6	0	0.0	29	60.4	44	91.7	0	0.0	4	8.3	28.860	<0.001*
Can the disease be cured?	30	62.5	0	0.0	18	37.5	46	95.8	0	0.0	2	4.2	16.168	<0.001*
What are the most prominent symptoms of phenylketonuria?	18	37.5	25	52.1	5	10.4	35	72.9	10	20.8	3	6.3	12.381	0.002*
What is the importance of treating the disease?	20	41.7	24	50.0	4	8.3	37	77.1	10	20.8	1	2.1	12.635	0.002*
What do you follow to treat phenylketonuria?	40	83.3	0	0.0	8	16.7	47	97.9	0	0.0	1	2.1	6.008	0.014*
How long is the treatment?	35	72.9	0	0.0	13	27.1	43	89.6	0	0.0	5	10.4	4.376	0.036*
When should treatment start?	36	75.0	0	0.0	12	25.0	45	93.8	0	0.0	3	6.3	6.400	0.011*
Is it possible to reach the normal level of intelligence when applying the correct diet from birth?	29	60.4	0	0.0	19	39.6	46	95.8	0	0.0	2	4.2	17.615	<0.001*
What is the probability that a parent with PKU will have a child with PKU again?	16	33.3	0	0.0	32	66.7	42	87.5	0	0.0	6	12.5	29.445	<0.001*
Can breast milk be given to a child with PKU?	25	52.1	0	0.0	23	47.9	39	81.3	0	0.0	9	18.8	9.188	0.002*
What foods should be avoided in phenylketonuria?	17	35.4	10	20.8	21	43.8	36	75.0	10	20.8	2	4.2	22.507	<0.001*
What protein alternatives are suitable for patients with phenylketonuria?	18	37.5	12	25.0	18	37.5	34	70.8	13	27.1	1	2.1	20.174	<0.001*
Mean±SD of total knowledge	12.65±2.51						24.09±2.75						21.287	<0.001*

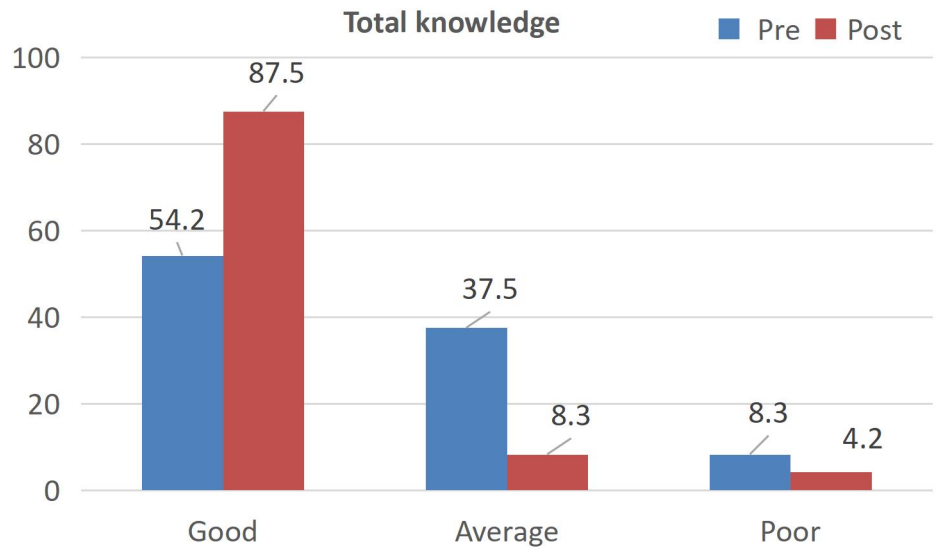

Figure (1): Distribution of Studied mothers regarding their Total Knowledge about PKU Disease and nutrition (n=48).

Table (3): Comparison of coping score pre- and posttest -intervention (n=48).

Items of coping scale	Pre				Post				Tests	
	Adequate		Inadequate		Adequate		Inadequate			
	N	%	N	%	N	%	N	%	t/X^2	P-value
Task-oriented	13	27.1	35	72.9	40	83.3	8	16.7	30.708	<0.001*
Emotional	16	33.3	32	66.7	37	77.1	11	22.9	18.577	<0.001*
Avoidant	14	29.2	34	70.8	36	75.0	12	25.0	20.202	<0.001*
Total coping	14	29.2	34	70.8	38	79.2	10	20.8	24.168	<0.001*
Mean±SD	79.8±7.2				193.43±9.26				67.116	<0.001*

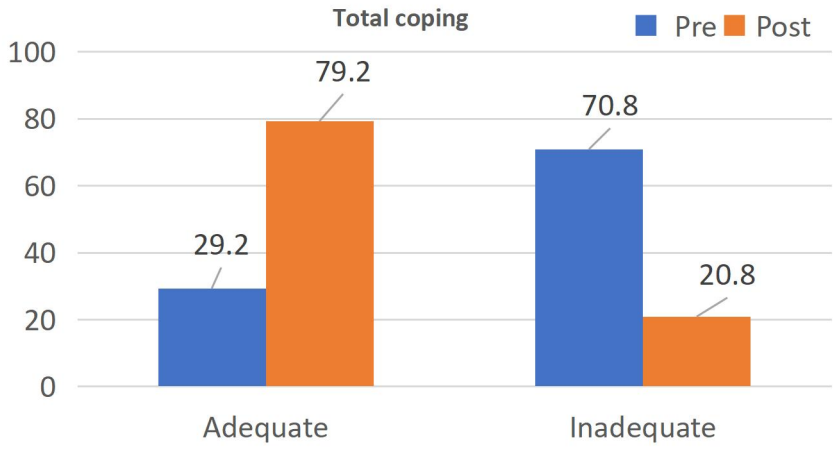

Figure (2): Distribution of total coping strategies of the Studied mothers (n=48).

Table (4): Comparison of total mother's strain levels pre- and posttest- coping strategies intervention (n=48).

Caregiver Strain Level	Pre		Post		Tests	
	N	%	N	%	t/X^2	P-value
High	35	72.9	12	25.0	22.051	<0.001*
Moderate	13	27.1	36	75.0		
Mean±SD	18.45±2.65		4.23±1.13		34.198	<0.001*

Table (5): Comparison of mother's quality-of-life domains (QOL) Pre- and Posttest- coping Strategies Intervention (n=48).

domains of QOL	Pre						Post						Chi-square	
	Good		Average		Poor		Good		Average		Poor			
	N	%	N	%	N	%	N	%	N	%	N	%	X^2	P-value
Physical Health	2	4.2	4	8.3	42	87.5	30	62.5	12	25.0	6	12.5	55.500	<0.001*
Psychological Health	5	10.4	8	16.7	35	72.9	33	68.8	11	22.9	4	8.3	45.746	<0.001*
Environment Health	9	18.8	6	12.5	33	68.8	34	70.8	9	18.8	5	10.4	35.766	<0.001*
Social Relations	4	8.3	7	14.6	37	77.1	32	66.7	9	18.8	7	14.6	42.482	<0.001*
Total QOL	5	10.4	7	14.6	36	75.0	31	64.6	10	20.8	7	14.6	38.865	<0.001*
Mean±SD	39.74±3.4						86.15±5.84						66.036	<0.001*

Table (6): Correlation between coping score and total knowledge pre- and post-intervention (n=48)

Items of coping	**Total Knowledge**			
	Pre		Post	
	r	P-value	r	P-value
Task-oriented	0.430	<0.001*	0.375	0.002*
Emotional coping	0.384	<0.001*	0.425	<0.001*
Avoidance coping	0.198	0.035*	0.169	0.015*
Total coping	0.427	<0.001*	0.372	<0.001*

Table (7): Correlation between caregiver strain, QOL Scores and coping score in pre- and posttest intervention (n=48).

	Caregiver Strain score		Coping score	
	r	P-value	r	P-value
Pre				
Coping score	-0.298	<0.001*		
QOL score	-0.234	0.003*	0.332	<0.001*
Post				
Coping score	-0.401	<0.001*		
QOL score	-0.352	<0.001*	0.463	<0.001*

Table (8): Relationship between some demographic data and coping strategies intervention among ~~the~~ mothers of children with PKU pre- and posttest intervention (n=48).

	Total coping											
	Pre						Post					
	Adequate		Inadequate		Chi-square		Adequate		Inadequate		Chi-square	
	N	%	N	%	X^2	P-value	N	%	N	%	X^2	P-value
Age												
27- <30	2	8.7	21	91.3	7.561	0.006*	9	39.1	14	60.9	4.703	0.030*
30 or more	11	44.0	14	56.0			22	88.0	3	12.0		
Level of education												
Can't read and write	0	0.0	3	100.0	9.379	0.025*	0	0.0	3	100.0	12.925	0.005*
Read and write	2	10.5	17	89.5			1	5.3	18	94.7		
Intermediate	5	31.3	11	68.8			3	18.8	13	81.3		
University	6	60.0	4	40.0			6	60.0	4	40.0		
Job												
Housewife	4	20.0	15	80.0	4.509	0.105	11	57.9	8	42.1	10.584	0.005*
working	10	34.5	19	65.5			27	93.1	2	6.9		

Discussion:

Mothers play a crucial role in caring for children with phenylketonuria (PKU). The responsibility of providing daily care for children may increase many burdens, leading to decreased quality of life and strain. Therefore, mothers must practice coping strategies that provides the ability and the flexibility to learn new skills and adapt to change with the burden of chronic disease which can help reduce the burden of strain and improve quality of life. The current study carried out to evaluate the effect of coping strategies intervention on strain and quality of life among mothers of children with phenylketonuria.

The results of the present study shows that less than quarter of mothers had higher education, more than half of mother's age≥ 30 years with mean of 31.04±5.41years and about two third of mothers were working and had insufficient income. This result is in accordance with a previous study carried out in Egypt **by Elsayed, Mohamed, AlRafay, & Khalifa, (2019),** who studied " Assessment of Mothers Care toward their Children having Phenylketonuria" revealed that the less than quarter of mothers had university education with their age were ranged from 20 to ≥ 40 years with a mean of 32.26 ± 4.2 years and two third of studied mothers had sufficient income. Furthermore, this results consistent with study of **Walkowiak et al., (2023),** who studied " Professional activity, gender and disease-related emotions: The impact on parents' experiences in caring for children with phenylketonuria" revealed that mean age of mothers were 37.8 years, range: 23–72 with less than half of them working full time. Additionally, **Öztürk, Bülbül, & Alpcan, (2022)**, who conducted a study about " Assessment of parents´ knowledge regarding phenylketonuria and its affecting factors: a cross-sectional study", in Turkey which reported that the mean ages of mothers were 34.53 ± 7.22, and less than two third of them had poor income.

The present study reveals that the majority of studied PKU' children are female with age ranged from 4 to less than 12 years with a mean of 8.54±2.46 years and more than one third of them had one sibling. Additionally, more than half of them had positive family history. From the researcher's point of view, in Egypt the consanguineous marriages are practiced for economic and social reasons which leads to the disease being hereditary between families. According to **Ahmed, (2017),** the prevalence of consanguineous marriages

was highest in rural Upper Egypt (43.6%) and lowest in urban Lower Egypt (13.2%). The previous finding is consistent with **by Elsayed, Mohamed, AlRafay, & Khalifa, (2019),** who reported that the majority of children with PKU were female (66.0%) with ranged age from birth to more than 9 years with a mean age of 4.45 ± 2.07 years and more than one third of them had one sibling (36.2%)

The present study findings demonstrate that mothers' knowledge of PKU had improved from before to after intervention, more than half of studied mother had good knowledge regarding PKU and diet of PKU at pre intervention. However, at post intervention the majority of studied mothers had good knowledge. From the researcher point of view, this result may be due to mothers' responsibility to provide daily care for their children and PKU is a lifelong condition and Adequate information about the child's PKU is important and helps the mothers cope with their desire to improve their knowledge to provide better care. This finding inconsistent with the study carried out by **by Elsayed, Mohamed, AlRafay, & Khalifa, (2019),** who reported that more than half of them have poor knowledge about PKU due to two third of mothers were housewife. This result was agreement with **Öztürk, Bülbül, & Alpcan, (2022)**, who reported that mothers had good knowledge about PKU and diet because they care for their children during visit and follow up in clinic and received training on PKU and its diet.

In relation to coping strategies, the present study findings revealed that less than three-quarters of the mothers use total coping strategies inadequate in pre intervention. After the intervention more than three quarter of mothers using total coping strategies adequately. Majority of mothers used task-oriented and three-quarters used emotional and avoidance coping. In the researcher's view, training mothers to practice strategies led to tolerance, reduction, and limitation of stressful events, which is necessary to reduce short-term strain before it turns into long-term stress and to cope with actual problems to solve them. This finding was supported by **by Elsayed, Mohamed, AlRafay, & Khalifa, (2019), ,** who reported that less than half of the mothers had poor total coping skills, the low percent of coping skill was related to that the researcher used only two scales from coping strategies as task – oriented and emotional coping. Similar to the study done by **Gheibizadeh, Gholami, Bassaknejad, Cheraghian, (2017),** who conducted a study about "Coping Strategies of Parents with Chronic Ill Children Hospitalized in educational hospitals" in Iran who found that majority of mothers use task-oriented and near three-quarters of mother used emotional strategies. Additionally, **Johnson & Onieka Mendoza, (2018),** who conducted a study about “Caregiving Coping Strategies Among Mothers with Chronically Ill Children" in India mentioned that, more than three quarters of mother used task – oriented and emotional coping.

The current study regarding mother’s strain levels, it shows that the mother had high strain level due to continuous stressful situation and care for chronic disease. Near three quarters of the mothers had high level of strain before intervention compared to one quarter after intervention with a highly statically significant difference between the two study phases. This outcome shows the effect of using the positive coping strategies intervention on mothers that decrease levels of strain. These results go with line **Johnson & Onieka Mendoza, (2018),** who reported that two third of mothers had high stress in pre intervention but after the program, stress was low as mothers practiced positive coping strategies to adapt with their chronically ill children. The presence of a child with chronic cause's continuous stress led to strain that effect on the family and especially the mothers. These results are in line with **Khairul, Kannan, & Erna, (2024**), who conducted a study about “The strain among caregivers of children with disabilities at the community-based rehabilitation centers in Kudat division of Sabah, Malaysia " who found that less than three-quarters of mothers had moderate strain and did not find any mothers had low strain due to continuous stress and tension.

In the present study regarding mother’s quality of life, it shows that three quarters of mothers had poor total quality of life especially, in physical, psychological and social relation before intervention compared to two- thirds of

them had good total quality of life in all dimension after intervention with there was a highly statistically significant difference between mothers' quality of life pre – post coping strategies intervention. Whereas mean of mothers 'quality of life in posttest was higher than pretest. From the researcher point of view, this improvement in quality of life of physical, psychological, environment and social relation due to mother's aware effective positive using of coping strategies intervention. This results agreement with **Irannejad, Dehghan & Mehdipour Rabori, (2018),** who conducted a study about "Stress and quality of life in parents of children with phenylketonuria" in Iran who found that the mean score of the quality of life of caregivers is lower (45.97) especially in physical and emotional health restriction and social relation due to They need for more time to carry responsibility (care, nourish, dress, medication, and cope with the child's sign and symptoms. As a result, mothers or caregivers of these children do not have enough time to deal with their needs and participate in social, relation, and activities, all of which affect the quality of life. These findings are consistent with a study by **Sikorová & Bužgová (2016),** who conducted a study about "associations between the quality of life of children with chronic diseases, their parents 'quality of life and family coping strategies" in outpatient clinic of faculty of medicine in Czechia who found that Parents of children with chronic diseases had lower quality of life in all domains due to problems with physical and psychosocial functioning tend to increase due to the presence of chronic disease, visits to outpatient clinics and continuous follow-up. These findings justified the research hypothesis.

The present study reveals a significant positive correlation between total knowledge and total coping scores both pre- and post-intervention, with task-oriented and emotional coping are stronger correlations than avoidance coping. These findings align with **Ibraheem, , Zaki, & Ahmad, (2016),** there is a statistically highly significant positive relation between knowledge and coping pattern with emotional coping and task oriented are positive correlation than avoidance coping. From research view, the mothers were understanding the importance of general coping strategies in influencing knowledge acquisition these results highlight that certain coping strategies, particularly task-oriented and emotional coping, play an important role in enhancing knowledge, suggesting that interventions aimed at improving these coping strategies lead to better knowledge.

The present study reveals a significant negative correlation ($P \leq 0.001$) between (caregiver stress and coping scores) and (caregiver stress and quality of life scores) in the pre- and post-test. Thus, mothers intervening to use coping strategies effectively leads to reduced levels of strain when facing difficult situations and continuous pressures due to their children's illnesses. Consequently, with adequate use of coping strategies, and reducing stress the quality of mothers' life was improved. This outcome aligns with the study conducted **by Irannejad, Dehghan & Mehdipour Rabori, (2018),** who found that there is a significant negative relationship between the overall scores of stresses and quality of life for mothers have children with PKU. From the researcher's point of view, the mothers of a children with PKU, when stress increases, quality of life decreases. Diagnosis of chronic illness for each child affects the quality of life of mothers. This outcome aligns with the study conducted **by Shaji Thomas, & Arulappan, (2021),** who conducted a study about "Health Related Quality of Life of Caregivers of Children and Adolescents With Phenylketonuria: A Systematic Review" who found that parents, particularly mothers, of children with PKU report lower health-related QoL scores compared to the general population and negative correlation between QOL and perceived stress has been reported. Therefore, it can be concluded that the QOL of parents decreases when the stress level increase Also, this outcome agreement with the study conducted by **Remor, Gabe, Teruya, & Doederlein Schwartz, (2024*)*** who conducted a study about "What is known about patients' quality of life with Phenylketonuria and their caregivers? A scoping review" who found that managing a child with PKU presents significant challenges that can negatively affect mothers' quality of life. While positively correlated with QOL and coping strategies intervention in the pretest and posttest with a highly statistically

significant positive correlation due to the mothers practiced and used coping strategies intervention led to improve the quality of life. These results go with line **Sikorová & Bužgová (2016),** who found that the strongest positive associations between coping strategies and parents' quality of life. These findings justified the research hypothesis.

The current study's findings showed significant correlation between the mothers' age, and coping strategies, the old mothers under 30 years showed markedly inadequate copings strategies in pre and still reporting lower coping abilities compared to those aged 30 or above in posttest intervention. Younger mothers had less life experience and less learning about coping strategies, making them more vulnerable to stress and less quality of life when managing a chronic condition like PKU. This finding aligns with **Elsayed, Mohamed, AlRafay, & Khalifa, (2019),** who found that younger mothers often experience lower coping strategy. According to educational levels, they were found mothers who could not read and write had inadequate using coping strategies pre and posttest intervention. The findings aligned with **Irannejad, Dehghan & Mehdipour Rabori, (2018)**, who found that the mothers with lower education level had low coping strategies than mothers with higher education levels. On the other hand, working mothers demonstrated significantly positive coping abilities pre and posttest intervention. These results are corroborated **by Elsayed, Mohamed, AlRafay, & Khalifa, (2019),,** who reported that a positive significant relation between mother's employee and coping strategies. From the researcher's point of view, employment may offer additional social support, financial stability, and opportunities for social engagement.

Conclusion

The study concluded that the coping strategies intervention was effective when applied by mothers who had children with phenylketonuria, which led to improve their quality of life and reduce their strain.

Recommendations

- Health education program about applying coping strategies for mothers with children phenylketonuria.
- Awareness of the health team teaches the mothers with children phenylketonuria how they practicing coping strategies.
- Design an educational book about coping strategies intervention for the mothers having children with PKU to help them enhancing quality of life and deal with strain situation.
- Further studies should be carried out on a large number of mothers with children phenylketonuria

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