Assessment of Adjustment Patterns of Children Suffering from Phenylketonuria

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Abstract

Background: Children with phenylketonuria necessitate adjusting and adapting their disease symptoms and lifestyle behaviors. **The study aimed to:** assess adjustment patterns of children suffering from phenylketonuria. **Setting:** This study was conducted at genetic clinic in outpatient department belonging to Children's Hospital affiliated to Ain Shams University Hospitals. **Design:** Descriptive research design was utilized. **Sample:** A purposive sample of 60 children attending with their mothers in the previously mentioned setting over a period of 6 months and were satisfying the inclusive criteria. **Tools:** Three tools were utilized in this study. The first tool was a structured questionnaire sheet to assess children' 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 three quarters of the studied children had unsatisfactory knowledge regarding phenylketonuria. Also, more than half of the studied children had moderate depression and severe level of anxiety. More than two thirds of the studied children had moderate loneliness and low self-esteem. There was an extremely statistically significant difference and positive correlation between children's total knowledge and total adjustment patterns. **Conclusion:** the minority of the studied children always adjusted with their disease. There was statistically significant difference between the studied children's characteristics and their total level of adjustment patterns. **Recommendations:** Continuous health educational programs about adjustment patterns should be provided for children suffering from phenylketonuria to cope positively with their disease.

Key words: Adjustment Patterns, Assessment, Children, Nursing, Phenylketonuria.

Introduction

Phenylketonuria (PKU) 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 **(Burton et al., 2018).**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 **(Elsayed et al., 2020).**

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 phenylacetate 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 **(Banta-Wright et al., 2016).**

A Phenylalanine restricted diet has been the mainstay treatment for PKU. Children treated early for PKU demonstrate subtle problems in cognitive function, school achievement, behavioral adjustment and quality of life. The current treatment for PKU involves strict metabolic control using a low-Phe diet of specialized medical foods. Small amounts of Phe from breast milk or commercial infant formula are considered sufficient for infants. In older children, the daily protein requirement is calculated, whereby a child is allocated a certain number of grams or units of daily protein. Foods such as eggs, milk, cheese, meat, poultry, fish, dried beans and legumes are excluded from the diet **(Dababneh et al., 2022).**

Children with PKU have behavioral problems including hyperactivity 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 children with PKU in a number of adjustment patterns (**Ashe et al., 2019**).

Pediatric nurses play a crucial role in assisting children with PKU 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 (**Carpenter et al., 2018**).

Aim of the Study

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

Research Questions:

1. What are the adjustment patterns of children suffering from phenylketonuria?
2. Is there relationship between adjustment patterns of children suffering from phenylketonuria and their characteristics?

Subjects and Methods:

Research Design:

A descriptive research design was utilized to achieve the aim of this study.

A. Research Setting:

The study was conducted at genetic clinic in outpatient department belonging to Children's Hospital affiliated to Ain Shams University Hospitals.

B. Subjects:

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

Inclusion Criteria:

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

Exclusion Criteria:
- Children with another chronic medical or mental health problem.

Tools of Data Collection:

Data were collected through using the following tools:

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: -

1) Characteristics of the children which include; age, gender, birth order, residence and history of the disease.
2) Characteristics of parents of children with phenylketonuria which include: age, level of education, occupation, monthly income and family history of the disease.

3) Knowledge of children regarding phenylketonuria which include; definition, causes, signs and symptoms, treatment, complications, prevention of phenylketonuria, prevention of complications and nutrition.

Questions were in the form of closed ended and multiple choices questions. The time consumed to fill in the questionnaire by the researcher for each child included in the study was 15-20 minutes. Mothers were questioned when necessary.

Scoring system for the studied children's knowledge about phenylketonuria:

A total score level for structured questionnaire sheet was 20 marks. The children's knowledge was checked with a model key answer sheet which prepared by the researcher and accordingly their total knowledge was categorized into either: satisfactory ($\geq 60\%$) or unsatisfactory ($< 60\%$).

Tool II: Psychometric Assessment

It was used to assess psychological problems of children with phenylketonuria through:

A) Children Depression Inventory (CDI):

Children Depression Inventory (CDI) was originally prepared by **Kovacs (1983),** and modified by **Abdel-Fattah,** (**1988)** 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.

Scoring system: each statement was answered by always, sometimes or never. The children were scored two degrees 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.

B) Children Manifest Anxiety Scale (CMAS):

Children Manifest Anxiety Scale (CMAS) by **Castaneda, (1965)** that was modified by **Abdel- Hamid and El-Nail, (1991)** 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 degree 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.

C) Children Loneliness Scale (CLA):

It was originally prepared by **Rasel, (1980)** and modified by **Al- Behery, (1985)** 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.

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.

D) Self – Esteem Inventory (SEI):

Self – Esteem Inventory (SEI) was developed by **Smith, (1967)** and modified by **El- Dosoki and Mosa, (1987)** 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 (14 items), school (11 items), social (11 items) and medical compliance (9 items). It included physical stressors (20 items), psychological stressors (14 items), social stressors (13 items) and financial stressors (10 items). 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:

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.

Pilot Study:

A pilot study was carried out on 6 children with phenylketonuria (10%) attending in the study 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 remodification 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 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 child with phenylketonuria and

accompanied mother individually for 20 to 30 minutes to gather the necessary data of the study.

III- 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.

Ethical Consideration

Verbal approval from parents and their children with phenylketonuria 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.

IV- 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. The relations between quantitative variables were tested by Pearson's correlation coefficient (r).

Results:

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

Variables	Number (No)	Percentage (%)
Age in years:		
6: < 8	5	8.3
8: < 10	10	16.7
10: ≤ 12	45	**75.0**
$\overline{X} \pm SD$	**10.16 ±1.51**	
Gender:		
Male	33	**55.0**
Female	27	45.0
Level of education:		
Not yet enrolled	2	3.3
Primary	48	**80.0**
Preparatory	10	16.7
Birth order:		
First	39	**65.0**
Middle	18	30.0
Last	3	5.0
Residence:		
Urban	14	23.3
Rural	46	**76.7**

Table (1) showed characteristics of the studied children. It revealed that three quarters of the studied children (75%) were aged 10: ≤ 12 years (X ± 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 child. It was cleared that more than three quarters of the studied children (76.7%) came from rural residence.

Table (2): Number and percentage distribution of the studied children according to their total level of knowledge regarding phenylketonuria (n=60)

Total level of children's knowledge	No	%
Satisfactory	15	25.0
Unsatisfactory	45	**75.0**

Table (2) revealed that three quarters of the studied children (75%) had unsatisfactory knowledge regarding phenylketonuria.

Table (3): Number and percentage distribution of the studied children according to their total level of depression (n=60)

Total level of depression	No	%
Mild (1: < 18)	20	33.3
Moderate (18: < 36)	35	**58.3**
Severe (36: ≤ 54)	5	8.3

Table (3) showed that more than half of the studied children (58.3%) had moderate depression.

Table (4): Number and percentage distribution of the studied children according to their total level of anxiety (n= 60)

Total level of anxiety	No	%
Mild (1: < 12)	7	11.7
Moderate (12: < 24)	23	38.3
Severe (24: ≤ 36)	30	**50.0**

Table (4) cleared that half of the studied children (50%) had severe anxiety.

Table (5): Number and percentage distribution of the studied children according to their total level of loneliness (n=60)

Total level of loneliness	No	%
Mild (1: < 13)	5	8.3
Moderate (13: < 26)	37	**61.7**
Severe (26: ≤ 40)	18	30.0

Table (5) represented that about two thirds of the studied children (61.7%) had moderate loneliness.

Table (6): Number and percentage distribution of the studied children according to their total level of self-esteem (n=60)

Total level of self esteem	No	%
Low (< 13)	40	**66.7**
Moderate (13: < 26)	14	23.3
High (26: ≤ 40)	6	10.0

Table (6) represented that more than two thirds of the studied children (66.7%) had low self-esteem.

Table (7): Number and percentage distribution of the studied children's total Adjustment patterns (n=60)

Adjustment patterns	Always		Sometimes		Never	
	No	%	No	%	No	%
Home	7	11.7	32	53.3	21	35.0
School	17	28.3	24	40.0	19	31.7
Social	7	11.7	29	48.3	24	40.0
Compliance with medication	10	16.7	20	33.3	30	50.0
Total	10	**16.7**	27	45.0	23	38.3

Table (7) showed that the minority of the studied children (16.7%) always adjusted.

Table (8): Relation between the studied children's total level of adjustment patterns and their characteristics (n= 60).

Children's characteristics	Total children's adjustment patterns						X^2	P Value
	Never		Sometimes		Always			
	No	%	No	%	No	%		
Age in years:								
6: <8	4	6.7	1	1.7	0	0.0		
8: <10	10	16.7	0	0.0	0	0.0	26.291	**0.000***
10: ≤12	9	15.0	26	43.2	10	16.7		
Gender:								
Male	13	21.7	13	21.7	7	11.7		
Female	10	16.7	14	23.2	3	5.0	15.443	**0.001***
Level of education:								
Not yet enrolled	1	1.7	1	1.7	0	0.0		
Primary	19	31.6	19	31.6	10	16.7	14.485	**0.006***
Preparatory	3	5.0	7	11.7	0	0.0		
Birth order:								
First	15	25.0	17	28.3	7	11.7		
Middle	8	13.3	9	15.0	1	1.7	11.405	**0.016***
Last	0	0.0	1	1.7	2	3.3		
Residence:								
Urban	7	11.7	6	10.0	1	1.7	11.661	**0.02***
Rural	16	26.6	21	35.0	9	15.0		

(*) Statistical significance difference, P <0.05

Table (8) showed that, there was statistically significant difference between the studied children's characteristics and their total level of adjustment patterns. It was observed that 43.2%, 21.7%, 31.6%, 28.3%, 35.0% and 25.0% of the studied children who were in the age group of 10: ≤12 years, males, in the primary stage, first child, came from rural residence were sometimes able to adjust.

Table (9): Correlation between the studied children's total knowledge and their total level of adjustment patterns.

Items	Total Children's Knowledge	
	r	**P value**
Total children's adjustment patterns	0.523	0.000*

(*) Statistical significance difference, P <0.05

Table (9) showed that, there was statistical significant difference and positive correlation between children's total knowledge and total adjustment patterns.

Discussion

In relation to the characteristics of the studied children, it was observed that, three quarters of the studied children were aged 10: ≤ 12 years ($\overline{X}$ ± SD 10.16 ±1.51 years). This study finding contradicted with a study conducted in Egypt by **Ibraheem et al., (2016),** entitled "Mother's Coping of Children Suffering from Phenylketonuria" who reported that, more than half of the studied children were aged 2: < 6 years.

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 and Elmoneem, (2016),** 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., (2020),** 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., (2020),** study which entitled "Assessment of Mothers Care toward their Children having Phenylketonuria"who found that, about half of the studied children were in primary school and more than one third of them 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., (2019),** 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.

Concerning the total knowledge of the studied children regarding to phenylketonuria, the current study showed that, three quarters of the studied children had unsatisfactory knowledge regarding phenylketonuria. This result was supported with **Fouad and Elmoneem, (2016),** who found that the majority of the studied sample had poor knowledge level regarding to 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.

Regarding to the total level of depression of the studied children, it was cleared that, more than half of the studied children had moderate depression, This result was supported with **Ford et al., (2018),** 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 children suffering from phenylketonuria perceived themselves as being different.

Regarding to the total level of anxiety of the studied children, it was noticed that, half of the studied children had severe anxiety. This result came in agreement with **Ashe et al., (2019),** 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. 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.

In relation to the total level of loneliness of the studied children, it was cleared that, about three quarters of the studied children always feel strange, isolated from others and social relationships are superficial. These findings are

similar to a study conducted by **Jahja et al., (2016),** 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 children suffering from phenylketonuria 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 that, more than two thirds of the studied children had low self –esteem. This result came in agreement with **Iakovou and Schulpis, (2020),** 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 children suffering from phenylketonuria always compare themselves with their peers in all aspects (physical, social and psychological) that may lead to a sense of inferiority.

As regards to total school adjustment of the studied children, It was noticed that only one third of the studied children able adjust to their school. This result came in agreement with **Ashe et al., (2019),** 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 children suffering from phenylketonuria feel different than others as a consequence of being ill. So, this feeling may prevent them from interaction with their peers and teachers.

In relation to total social adjustment of the studied children, it was cleared that, minority of the studied children always socially adjusted. This result was supported with **Ford et al., (2018),** who found that most of children with PKU withdrew from social situations and became socially isolated.

Concerning to the relation between the studied children's total level of adjustment patterns and their characteristics, the current study 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: ≤12 years, males, in the primary stage, first child, came from rural residence were sometimes able to adjust.

Conclusion:

The present study concluded that the minority of the studied children always adjusted with their disease. There was statistically significant difference between the studied children's characteristics and their total level of adjustment patterns.

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.

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