

# (uHFW RI QXUVLQJ LQWHUYHQWLRQ RQ DGMXV IURP SKHQ\ONHWRQXULD

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%DFNJURXQG 3KHQ\ONHWRQXULD LV DQ LQKHULWHG GLVRUGHU FDOOHG SKHQ\ODODQLQH LQ WKH EORRG

\$LP ,PSURYH DGMXVWPHQW SDWWHUQV RI FKLOGUHQ VXuHULQJ I 0HWKRGRORJ\ 7KLV VWXG\ ZDV FRQGXFWHG DW JHQHWLF FOLQL FKLOGUHQ¶V KRVSLEWDO DVOLDWHG WR \$LQ 6KDPV XQLYHUVLW\ K GHVLJQ ZDV XWLOLJHG

\$ SXUSRVLYH VDP SOH RI FKLOGUHQ DFFRPSDQLHG E\ WKHLU P VHWWLQJ RYHU D SHULRG RI PRQWKV DQG VDWLVI\LQJ WKH LQF 7KHU WRROV ZHUH XWLOLJHG LQ WKLV VWXG\ SUH SRVW QXUVLQJ TXHVWLRQQDLUH VKHHW WR DVVHVV FKLOGUHQ¶V NQRZOHGJH UHJD SV\FKRPHWULF DVVHVVPHQW WR DVVHVV SV\FKRORJLFDO SUREOH WKLUG WRRO ZDV DGMXVWPHQW SDWWHUQV VFDOH RI FKLOGUHQ 5HVXOWV 7KH VWXG\ ; QGLQJ UHYHDOHG WKDW WKHUH ZDV VWD VWXGLHG FKLOGUHQ¶V NQRZOHGJH SUH DQG SRVW QXUVLQJ LQV VWDWLWLFDO VLJQL; FDQFH GLuHUHQFH UHJDUGLQJ WR WRWDO SUH DQG SRVW QXUVLQJ LQWHUYHQWLRQ LPSOHPHQWDWLRQ \$ VLJQL; FDQW GLuHUHQFH DQG SRVLWLYH FRUOHODWLRQ EHWZHH DGMXVWPHQW SDWWHUQV SUH DQG SRVW QXUVLQJ LQWHUYHQWL &RQFOXVLRQ 7KHUH ZDV WKHUH ZDV D SRVLWLYH HuHFW RI WKH SDWWHUQV RI FKLOGUHQ VXuHULQJ IURP SKHQ\ONHWRQXULD

5HFRPPHQGDWLRQV &RQLQRRXV KHDOWK HGXFDWLRQDO SUR DVVHVVPHQW DERXW DGMXVWPHQW SDWWHUQV VKRXOG EH SU SKHQ\ONHWRQXULD WR FRSH HuHFWLYHO\ ZLWK WKHLU GLVHDVH

. H\ZR UAGJvstment Patterns, Children, Intervention, Program, Phenylketonuria, Nursing.

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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 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 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

**Research hypothesis**

Children with phenylketonuria have lower scores on the Children Depression Inventory (CDI) compared to children with other genetic disorders.

**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 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**

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.

The researcher after reviewing the current available literature and 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

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

was used to assess the psychological problems of children with phenylketonuria through.

Children Depression Inventory (CDI): Children Depression Inventory (CDI) was originally prepared by Kovacs, and 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 1-10 referred to mild depression, score 11-15 referred to moderate depression, and score 16-54 referred to severe depression.

Children Manifest Anxiety Scale (CMAS): Children Manifest Anxiety Scale (CMAS) 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 1-10 referred to mild anxiety symptoms, score 11-20 referred to moderate anxiety symptoms, and score 21-36 referred to severe anxiety symptoms.

Children Loneliness Scale (CLA): It was originally prepared by Reilly et al. [3] to assess the degree of severity of loneliness symptoms of children. 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 1-10 referred to mild loneliness symptoms, score 11-20 referred to moderate loneliness symptoms, and score 21-40 referred to severe loneliness symptoms.

Self-Esteem Inventory (SEI): Self-Esteem Inventory (SEI) [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



9DULDEOHV	1XPEHU 1R	3HUFHQWDJH
\$JH LQ \H DU V		
	5	
	10	16.7
"	45	75
X ± SD	10.16 ± 1.51	Range
* H Q G H U		
Male	33	55
Female	27	45
/HYHO RI HG XFDWLRQ		
Not yet enrolled	2	3.3
Primary		
Preparatory	10	16.7
% L U W K R U G H U		
First	39	65
Middle		30
Last	3	5
5 H V L G H Q F H		
Urban	14	23.3
Rural	46	76.7

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

9DULDEOHV	1R	
&KLOG↑V DJH DW GLDJQ \H DU V	RVLV RI WKH GLVH DVH LQ	
<3		46.7
3:<6	16	26.7
6:<9	12	20
"	4	6.7
7KH GLVH DVH ZDV GLVFRYHUHG E\		
Chance	11	
Signs and symptoms of the disease	12	20
Investigations	30	50
Complications of the disease	7	11.7
1XPEHU RI IROORZ XS SHU PRQWK		
Once	41	
Twice	19	31.7

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

9DULDEOHV	0RWKHU		)DWKHU	
	1R		1R	
	Age in years			
'	6	10	3	5
'		63.3	20	33.3
"	16	26.7	37	61.7
X ± SD	32.90 ± 6.17	"	30	30
/HYHO RI HG XFDWLRQ				
Illiterate	6	10	6	10
Read and write	3	5	2	3.3
% D V L F H G X F D W L R Q		15	7	11.7
Technical education	30	50	31	51.7
High education	12	20	14	23.3
2 F F X S D W L R Q				
Working	14	23.3	52	
Not working	46	76.7		13.3

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



7RWDO O ORQHOL	HYHO RI 3UH		3RVW		X <sup>2</sup>	S YDOXH
	1R		1R			
0LOG	5				14.645	0.001**
0RGHUDWH	37	61.7	12	20		
6HYHUH	"	30	0	0		

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

7RWDO O HVWHHP	HYHO RI VHOI		3RVW		X <sup>2</sup>	S YDOXH
	1R		1R			
/RZ	40	66.7	0	0		
0RGHUDWH	14	23.3	17			
+LJK	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).

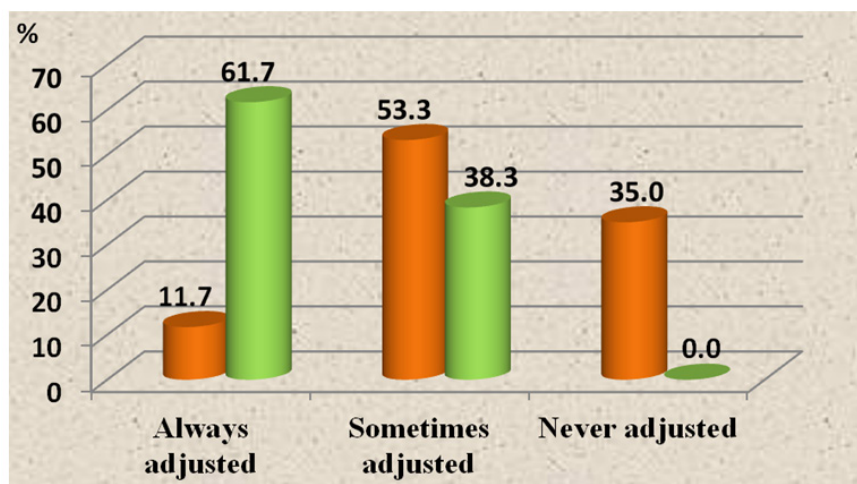


Figure 1. Percentage distribution of the studied children’s total home adjustment pre/post nursing intervention (n=60). Note: Pre-intervention: (orange); Post-intervention: (green).

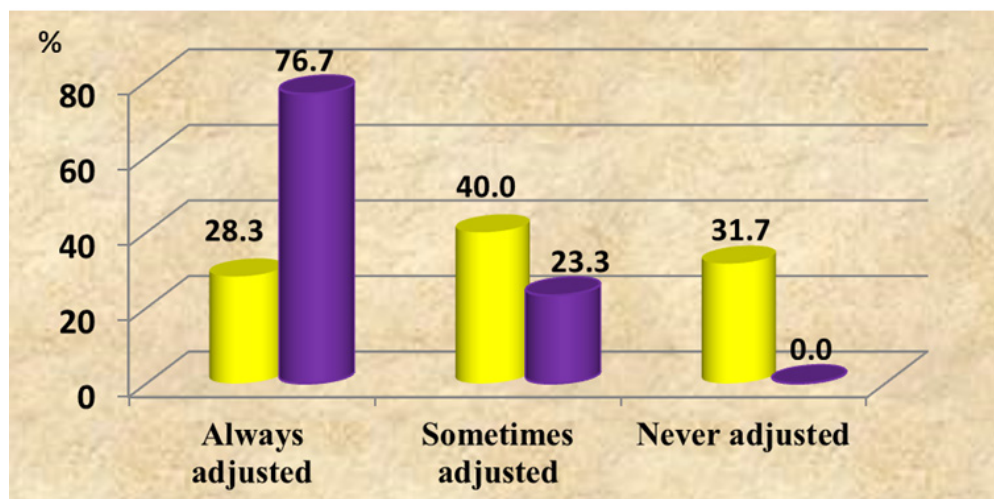


Figure 2. Percentage distribution of the studied children’s total school adjustment pre/post nursing intervention (n=60). Note: Pre-intervention: (yellow); Post-intervention: (purple).

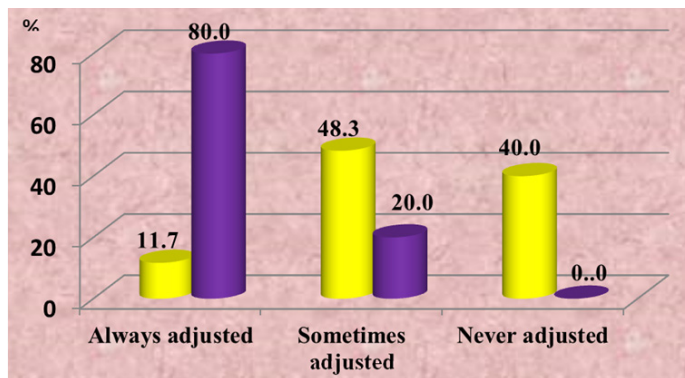


Figure 3. Percentage distribution of the studied children’s total social adjustment pre/post nursing intervention (n=60). Note: Pre-intervention: ( ■ ); Post-intervention: ( ■ ).

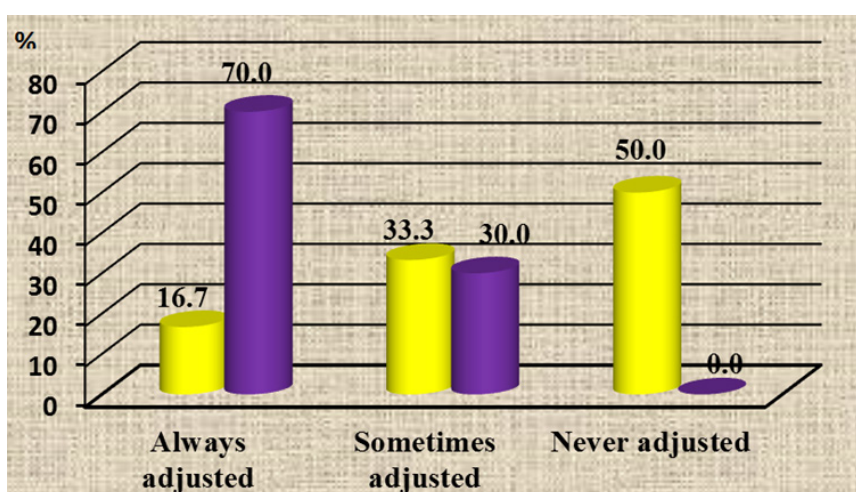


Figure 4. Percentage distribution of the studied children’s total compliance with their medication pre/post nursing intervention (n=60). Note: Pre-intervention: ( ■ ); Post-intervention: ( ■ ).

7 R W D O O H Y H V W H H P	H O R I V H O I H		3 R V W		X <sup>2</sup>	S Y D O X H
	1 R		1 R			
Always adjusted	10	16.7	43	71.7		0.003
Sometimes adjusted	27	45	17			
Never adjusted	23		0	0		

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

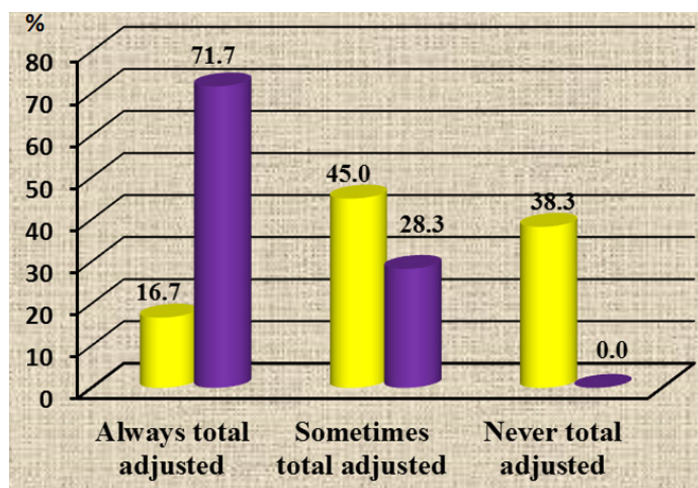


Figure 5. Percentage distribution of the studied children’s total adjustment patterns pre/post nursing intervention (n=60). Note: Pre-intervention: ( ■ ); Post-intervention: ( ■ ).

Total children's adjustment patterns												
3UH						3RVW						
&KLOGUHQTV FKDUDFWHULVWLFV	1R		1R		X <sup>2</sup>	S Y D O X H	1R		1R		X <sup>2</sup>	S Y D O X H
	1R	1R	1R	1R			1R	1R	1R	1R		
Age in years	4	6.7	1	1.7	0	0	0	0	5	0	0	0
	10	16.7	0	0	0	0.000**	0	0	13.3	2	3.3	34.2
"	9	15	26	43	10	16.7	0	0	4	41	0	0.000**
Gender												
Male	13	21.7	13	22	7	11.7	0	0	11	22	36.7	0.003
Female	10	16.7	14	23	3	5	0	0	6	21	35	
Level of education												
Not yet enrolled	1	1.7	1	1.7	0	0	0	0	2	3.3	0	0
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
%LUWK RUGHU												
First	15	25	17		7	11.7	0	0	12	20	27	45
Middle		13.3	9	15	1	1.7	0	0	5		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	0	0	3	5	11	
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	0	0	5		23	
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
"	2	3.3	2	3.3	0	0	0	0	0	0	0	0
The disease was discovered by												
Chance	5		4	6.7	2	3.3	0	0	4	6.7	7	11.7
Signs and symptoms	7	11.7	4	6.7	1	1.7	0	0	5		7	11.7
Investigations	9	15	16	26.7	5		0	0	7	11.7	23	36.7
Complications	2	3.3	3	5	2	3.3	0	0	1	1.7	6	10
Number of follow up/month												
Once	16	26.7	16	26.7	9	15	0	0	11		30	50
Twice	7	11.6	11		1	1.7	0	0	6	10	13	21.7

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







complications.

• Further researches are required involving larger study sample knowledge, attitudes, and practices and empowering the adjustment patterns positively.

( W K L F D O & R Q V L G H U D W L R Q V

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. Their privacy and confidentiality were guaranteed.

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9. & DVWDQHGD \$ % R \ G 0 F & DQG O H V F a c t o r s o f c h i l d r e n ' s f r o m o f m a n i f e s t a n x i e t y s c a l e : S o c i e t y f o r r e s e a r c h i n c h i l d d e v e l o p m e n t 1 9 5 6 ; 2 7 ( 3 ) : 3 1 7 - 3 2 6 .
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