Assessment of Lifestyle for Children with Phenylketonuria

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Abstract

Background: Phenylketonuria is an autosomal recessive disorder characterized by accumulation of phenylalanine in blood and body fluids that is caused by defective Phe hydroxylase activity. **Aim** of the present study is to assess lifestyle for children with phenylketonuria. **Design:** A descriptive design was used in this study. **Setting**: conducted at genetic counseling clinic in basic health care unit at El Fayoum affiliated to Ministry of Health (MOH). **Sample:** A purposive sample included all children with Phenylketonuria composed of 79 children accompanied by their mothers. **Tools:** structural interviewing questionnaire to assess demographic characteristics, and medical history, and lifestyle assessment Questionnaire for children with PKU to identify current strengths of child's health, and risk factors. **Results:** (74.7%) of families were discovered the disease by Ministry of Health screening programs. (92.4%) of children were made regular follow up. (78.5%) of children had follow up visits once monthly. 89.9% of children were not Taking breast feeding 62% of mothers had inadequate knowledge about the relation between breast feeding and the disease process. **Conclusion:** There is a statistically significant correlation between compliance with the prescribed diet and growth problems. There is no statistically significant correlation between compliance with the prescribed diet and sleep problems. **Recommendations:** The current study recommended Periodic assessment of Children affected commitment to prescribed diet. Increase mother knowledge about the importance of prescribed diet for children with Phenylketonuria.

Key words: life style, children, Phenylketonuria.

Introduction

Phenylketonuria (PKU) is an autosomal disorder characterized by accumulation of phenylalanine (Phe) in blood and body fluids that is caused by defective Phe hydroxylase activity. PAH deficiency is classified as classical PKU (Phe N 1200 μmol/L), mild PKU (Phe 600–≤1200 μmol/L) or mild hyperphenylalaninaemia (Phe b 600 μmol/L). The incidence of PKU varies according to ethnic background. In the UK, PKU affects about 1 every 10,000 newborns of white European ancestry with around 70 babies born with PKU annually, suggesting that over 6000 people in the UK have PKU ***(Williams, Mamotte, Burnett, 2018).***

Provisional diagnosis to be Metachromatic Leukodystrophy (MLD)/ Aminoaciduria/ Leigh syndrome, advising for urine and serum amino acid test, thyroid hormone stimulating test, creatinine phosphor- kinase (CPK), nerve conduction study (NCS), serum lactate pyruvate tests. Physiotherapy and on anti-

epileptic medications. Later when the child visited for the outpatient department with conditions of global developmental delay, right focal seizures, poor visual perception and not yet attained social smile, head control, and rolling eyes during the 2nd month after birth ***(MacDonald, 2017).***

On examination weight 7 kg, hypo pigmented scalp hair, increased tone, deep tendon reflex (DTR) scoring 5.MRI-brain suggests of severe global developmental delay/microcephaly/seizure disorder. Screening for plasma amino acid analysis is recommended using reverse phase HPLC. The treatment continued as the same. The diagnosis was confirmed by elevated levels of phenylalanine levels to 300 µmol/L (31-75 µmol/L), urine metabolic screening reports are positive for ferric chloride test. Recommending for Phenylalanine restricted diet to be initiated, also kept on regular anti-seizure medications and recommended regular physiotherapy ***(The National Society for Phenylketonuria United Kingdom, 2016).***

If the child Left untreated, with prolonged high Phe concentrations in the blood classical PKU could result in severe cognitive impairment, seizures, behavioral problems and features of autism. Such irreversible complications could be avoided through early treatment with a low-Phe diet from the first few weeks after diagnosis and throughout life. Pharmacological treatments are also available and currently include the synthetic formulation of tetrahydrobiopterin (BH4), sapropterin dihydrochloride, and medical food or amino acid supplements, which including formulations of large neutral amino acids such as tryptophan and tyrosine ***(Hanley, 2017).***

Blood tests should conduct by parents at home usually with the following frequency: weekly for children aged ≤5 years old and between twice monthly and monthly for children ≥6 years. Thus, many aspects of PKU management appear as time-consuming for caregivers who manage the PKU lifestyle on top of regular childcare. PKU has already been shown to affect caregiver's life and even their ability to continue regular work responsibilities, with 11% of parental caregivers in one of the survey reporting that they had to stop paid work and 20% reporting that they had to change their jobs to meet the child's needs ***(Loeber, 2017).***

Adherence to this treatment regimen is considered Demanding as they need to purchasing special types of foods, children supervising the intake of Phe-free L-amino acid supplements planning daily Phe consumption, preparing low-Phe meals (often involving additional cooking), monitoring Phe intake, and attending clinic visits ***(Hanley, 2017).***

Dietary treatment of PKU is achieved via a low-Phe diet that severely restricts the intake of natural protein in order to achieve control of blood Phe concentrations. The low-Phe diet includes food that is natural. Low in protein, such as fruit, some vegetables, fats and oils, as well as specially formulated low-protein products, such as low-protein flour, pastas and bread. All patients on dietary treatment require Phe-free L-amino acids, usually supplemented with additional carbohydrate, with or without fat, vitamins and minerals. Adherence to dietary treatment is essential, thus careful planning, dietary supervision and monitoring by the care giver are required ***(Blau, van Spronsen, Levy, 2018).***

Parental Quality of Life (PQoL) may be the most appropriate indicator of

parental adjustment. It is defined by physical and psychological well-being and social functioning and covers health-related dimensions like "physical and daily functioning", as well as dimensions referring specifically to the chronic condition of their child, like "satisfaction with the family", and "emotional stability". PQoL differs from the construct of health-related quality of life (HRQoL) in its explicit relation to the chronic condition of the child ***(Fidika, 2019).***

Additionally, assessing PQoL through using psychometric measure enables the identification of parents with sub-clinical impairments due to the chronic condition of their child. QoL of parents having an ill children comparing of parents having healthy schoolchildren showed that almost half of the parents of chronically ill children are at risk of impairment in their QoL. Impairment in QoL among parents with chronically ill children, particularly among parents having children with heart disease ***(Quittner, Glueckauf, Jackson, 2016).***

Significance of the study

Phenylketonuria constitute a major health problem throughout child's life and their family when dangerous build up occur for a person with Phenylketonuria who eat foods high in protein, as the gene defect is unknow passed down from generation to generation ***(Anderson & Leuzzi, 2014).*** PKU still has adverse effects on children in Upper Egypt leading to developmental problems, as mental retardation, convulsions, delayed social and intellectual skills, delayed growth of weight and length, bad odor of the body, skin rash and eczema with behavioral abnormalities ***(Abdel Rahim et al., 2013).***

The incidence of PKU varies widely in different human populations. These disorders are frequent affect males and females. In Egypt the incidence of PKU is unknown, but all cases about 8000 cases follow up in genetic clinics in Egypt. They are diagnosed through performing screening test of serum Phe level between one and seven days after birth, through pricking the heel of the newborn and test the blood obtained for phenylalanine concentration ***(Georgianne, 2014).***

Aim of the study

This study aimed to assess lifestyle for children with phenylketonuria.

Research Question:

What is current lifestyle for children with phenylketonuria?

Subjects and Methods

The subject and methods of the current study were discussed under the following four designs:

I. Technical Design
II. Operational Design
III. Administrative Design
IV. Statistical Design

I- Technical Design:

The technical design for the study included research design, setting of the study, study subjects, and tools of data collection.

Research design:

A descriptive research design was used to assess lifestyle for children with phenylketonuria.

Research Settings:

The current study was conducted at genetic counseling clinic in basic health care unit in Fayoum affiliated to MOH.

Research Subjects:

A purposive sample that included all children with PKU composed of 79 child accompanied by their mothers who were attend to the previously mentioned setting according to the following criteria; Children aged from one year to 5 years old and free from other medical diseases such as (DM, Heart diseases, Hepatitis B).

Tools of data collection:

Two data collection tools were used to carry out the current study namely: **An Interviewing Questionnaire:** It was designed by the researcher after reviewing related literature and translated into Arabic language it consisted of two parts as the following:

Part I: It concerned with demographic characteristics of child, such as age, gender, and child ranking. Socio-demographic characteristics of mothers regarding age, gender, relative degree, marital status, educational level, occupation, and family monthly income.

Part II: It related to medical history of children with PKU, including duration, discovery of the disease, follow up frequency per month, the degree of follow up, history of family members. And the relationship of family member suffering from the disease, complications of the disease and measurement of the child (Hight, weight, body mass index and phe serum level).

Lifestyle Assessment Questionnaire for children with PKU: It was adapted from ***Naturopathic foundation health clinic, (2014)*** and used to identify current strengths of child's health, any risk factors that might be present, and highlight recommendations that may want to consider. It consisted of nine categories, general information, mothers' health during pregnancy, family history and information, first few years of child life, past and present health concerns, general information on diet, atypical day for child, child patterns of behavior, review of the child physical system.

❖ **Scoring system**

For each of the knowledge items, a correct response was scored "one", and an incorrect "zero". For each area of knowledge, it was considered satisfactory if the percent score was 60% or more and unsatisfactory if less than 60%.

II- Operational Design:

The operational design for this study included three phases namely; preparatory phase, pilot study, and field work.

Preparatory phase:

This phase started with a review of the current and past, national and international related literature concerning the subjects of the study, using textbooks, articles, journals, and websites. This review was helpful to the researcher in reviewing and developing the data collection tools, and then the researcher tested the validity of the tool through jury of expertise to test the content, clarity, accuracy, and relevance of questions for tools.

Tools Validity and Reliability:

Face and content validity of the study tools was assessed by jury group consisted of three (Professors) in pediatric nursing. Jury group members judge the tools for comprehensiveness, accuracy and clarity in language. Based on their recommendation's correction, addition and / or omission of some items were done. The study tools were tested for its internal consistency by Cronbach's Alpha. It was 0.783 for the Interviewing questionnaire and 0.815 for lifestyle assessment Questionnaire for children with PKU.

Pilot study:

A Pilot study was carried out on 10% of the total study sample (eight children) to evaluate the applicability, efficiency, clarity of tools, assessment of feasibility of field work, beside to detect any possible obstacles that might face the researcher and interfere with data collection. Necessary modifications were done based on the pilot study findings such as (omission of some questions from tool) in order to strengthen their contents or for more simplicity and clarity. The pilot sample was excluded from the main study sample.

Field work:

Data collection of the study was started at the beginning of October 2018, and completed by the end of April 2019. The researcher attended at genetic counseling clinic in the basic health care unit two days per week from 9am to 2pm. The researcher first explained the aim of the study to the mothers and reassures them that information collected will be treated confidentiality and that it will be used only for the purpose of the research.

III- Administrative Design

An official letter requesting permission to conduct the study was directed from the dean of the faculty of nursing Ain Shams University to the directors of genetic counseling clinic in basic health care unit in El Fayoum affiliated to MOH to obtain their approval to carry out this study. This letter included the aim the study and photocopy from data collection tools in order to get their permission and help for collection of data.

Ethical Consideration

Prior the study conduction, ethical approval was obtained from the scientific research ethical committee of the faculty of nursing, Ain Shams University. The researcher met director of the genetic counseling clinic in basic health care unit in Fayoum affiliated to MOH to clarify the aim of the study and take their approval. The researcher also met the mothers to explain the purpose of the study and obtain their approval to participate in the study. They were reassured about the anonymity and confidentiality of the collected data, which was used only for the purpose of scientific research. The subjects' right to withdraw from the study at any time was assured.

IV- Statistical Design:

The collected data were coded and entered into the statistical package for the social science (SPSS 23.0). Data was presented and suitable analysis was done according to the type of data obtained for each parameter. Data were presented using descriptive statistics in the form of frequencies and percentages for categorical variables, and means and standard deviations for continuous

quantitative variables. Qualitative categorical variables were compared using Chi-square (X^2) test. Person and spearman correlation was used to examine the correlation between quantitative and qualitative variables. Statistical significance was considered when P-value < 0.05.

system. All of mothers prepared food for their children. less than three quarter of children (73.4%) were taken diet system free from protein. more than two thirds of children (69.6%) were Compliance with the diet system. Majority of children (84.8%) were dispensing the prescribed diet once every month. Majority of children (88.6%) were dispensed amount of the prescribed diet. Majority of children's mothers (89.9%) were Using calculation method to measure amount of foods, and less than half of children's mothers (40.7%) calculate amount of foods from the metabolic clinic. Majority of mothers (88.6%) were keeping the prescribed diet out of refrigerator.

Table (5): Shows that less than three quarters of children (74.7%) not enrolled to nursery and more than two fifth of them (41.8%) reported the main cause is late speech development for their children. About one quarter of them (25.3%) didn't able to cope due to fear of others in the nursery. The main cause of refusing taking foods in the nursery is Unawareness of the teacher to the importance of eating the prescribed foods.

Table (6): Shows that there is a highly statistically significant correlation between compliance with the prescribed diet and effect of the disease on general health.

Results

Table (1): Show that one third of children's weight (31.6%) were 9:<11 kg, one third of children's height (31.6%) were 75:<85 cm. less than half of children (45.6%) were normal in body mass index.

Table (2): Show that disease affected on less than half (43%) of children's general health such as; Delayed growth of weight and length, Behavioral problems, delayed social and intellectual skills, Skin related disease, Convulsions, and Bad odor of the body. majority of children (87.3%) had no complication of the disease. All children (100%) were made Phenylalanine level in the blood as regular investigation. Less than half of children (49.4%) made Phenylalanine level in the blood once every two months.

Table (3): Show that more than half of children (50.6%) were taking solid food after one year. Less than two thirds of children (63.3%) were taking three meals per day and more than half (55.7%) of the largest meal was lunch. Less than half (43%) of preferred food for children were Carbohydrates (rice – bead). More than half of preferred foods for children (59.5%) were prescribed diet. More than three quarter of children (78.5) took the last meal from 6pm to 10pm. More than two thirds of children (72.2%) were receiving adequate fluids and more than three fifth of children (62%) were receiving Permitted fluids (water – low sugar fruits).

Table (4): Show that majority of children (81%) had followed special diet

Table (1): Distribution of the studied children According to their Physical characteristics (n=79).

Physical characteristics	No.	%
Child weight		
9:<11 kg	25	31.6
11:<13 kg	16	20.3
13:<15 kg	20	25.3
15:<17 kg	12	15.2
>17 kg	6	7.6
Child length		
<75 cm	4	5.1
75:<85 cm	25	31.6
85:<95 cm	11	13.9
95:<100 cm	19	24.1
100:<107 cm	16	20.3
> 107 cm	4	5.1
Body mass index		
Normal	36	45.6
Less than normal	20	25.3
More than normal	11	13.9
Obese	12	15.2

Table (2): Distribution of the studied children according to effect of the disease on their general health (n=79).

Items	No.	%
Effect of the disease on general health		
Yes	34	43
No	45	57
Effect of the disease on general health		
Delayed growth of weight and length	0	0
Behavioral problems	8	10.1
Delayed social and intellectual skills	12	15.2
Skin Turger	0	0
Convulsions	0	0
Bad odor of the body	0	0
All of the above	14	17.7
Presence of complications of the disease		
Yes	10	12.7
No	69	87.3
Complications		
Mental retardation	0	0
Behavioral problems	6	5.1
Delayed social and intellectual skills	4	7.6
Convulsions	0	0
Head larger than normal	0	0
Head smaller than normal	0	0
Skin rash and eczema	0	0
Types of regular investigations		
Phenylalanine level in the blood	79	100
Urine analysis	0	0
CBC	0	0
All of the above	0	0
Number of investigating Phenylalanine level in the blood		
Once monthly	36	45.6
Once every two months	39	49.4
Once every three months	4	5

Table (3): Distribution of the studied children according to their nutrition life style (n=79).

Items	No.	%
Age of child when taking solid feeding		
< one year	39	49.4
≥ one year	40	50.6
Number of the main meals per day		
Two meals	29	36.7
Three meals and more	50	63.3
Time of the largest meal		
Breakfast	15	19
Lunch	44	55.7
Dinner	20	25.3
Preferred food for children		
Carbohydrates (rice – bead)	34	43
Protein (meat – chicken)	23	29.1
Vegetables and fruits	9	11.4
The described diet	13	16.5
Un-preferred food for children		
Carbohydrates (rice – bead)	10	12.7
Protein (meat – chicken)	12	15.2
Vegetables and fruits	10	12.7
The described diet	47	59.4
Last meal in the day		
From 6pm to 10pm	62	78.5
After 10pm	17	21.5
Receiving adequate fluids		
Yes	57	72.2
No	22	27.8
Types of fluids		
Permitted fluids (water – low sugar fruits)	49	62
Not permitted fluids (canned juices)	8	10.1

Table (4): Distribution of the studied children according to their prescribed diet system (n=79).

Items	No.	%
Child follow special diet system		
Yes	64	81
No	15	19
Foods prepared by:		
Physician or nurse	0	0
Dietitian	0	0
The mother	79	100
Type of diet system		
Free from protein	58	73.4
Free from Fenyl-Alanin	17	21.5
Low fat	4	5.1
Compliance with the diet system		
Yes	55	69.6
No	24	30.4
Cause of no compliance with the diet system		
The child don't like it	21	26.6

Items	No.	%
High cost of diet	3	3.8
Number of dispensing the prescribed diet during the month		
Once	67	84.8
Twice	8	10.1
More than two times	4	5.1
Adequacy of the dispensed amount of the prescribed diet		
Yes	70	88.6
No	9	11.4
Dealing to compensate in adequacy		
Give the child the permitted vegetables and fruits	9	11.4
Calculation method to measure amount of foods		
Yes	71	89.9
No	8	10.1
Type of calculation method of measuring amount of foods		
From the metabolic clinic	40	40.7
15 cup for every 8 meals	31	39.2
Place of keeping the prescribed diet		
In the freezer	3	3.8
Out of refrigerator	70	88.6
In special place in the refrigerator	6	7.6

Table (5): Distribution of the studied children according to their types of education(n=79).

Items	No.	%
Child enrollment to nursery		
Yes	20	25.3
No	59	74.7
Reason for not enrolled to nursery		
Late of speech	33	41.8
Small age	12	15.2
Dependency of child	14	17.7
Age of enrollment to nursery		
1:<3 years	5	6.4
3:< 5 years	15	18.9
Adapting in the nursery		
Yes	12	15.2
No	8	10.1
Reason for not adapted in the nursery		
Fear of others	20	25.3
Presence of education or comprehension problems		
Yes	8	10.1
No	12	15.2
Types of education or comprehension problems		
Inability to follow instruction	20	25.3
Inability to calculate mathematic issues	0	0
Difficulty of finishing tasks	0	0
Difficulty of concentration	0	0
Normal social behavior		
Yes	16	20.2
No	4	5.1
Types of social behavior problems		
Violence against other children	16	20.2
Take belonging of other children	4	5.1
Taking the prescribed foods in the nursery		
Yes	12	15.2
No	8	10.1
Reason of didn't taking their food		
Child desire to take other children's foods	12	15.2
Unawareness of the teacher to the importance of eating the prescribed foods	8	10.1

Table (6): Correlation between compliance with the prescribed diet and effect of the disease on general health

Variables		Compliance with the prescribed diet
Effect on general health	R	0.784
	P	0.000**

(**) Highly statistically significant correlation at P-value <0.05

Discussion

Regarding Physical characteristics of the studied children, the current study revealed that one third of children's weight were 9:<11 kg, one third of children's height were 75:<85 cm. nearly half of children were normal in body mass index.

This result is in congruence with the study done by **MacDonald et al. (2016)** who studied "The personal burden for caregivers of children with phenylketonuria" a cross-sectional study investigating time burden and costs in the UK. Molecular genetics and metabolism reports and found that majority of the participants reported that majority of children's weight were 9:<13 kg, and half of children were normal in body mass index. This result is in agreement with the study done by **Al-Zyoud et al. (2019)** who studied "Culturable gut bacteria lack Escherichia coli in children with phenylketonuria" and found that half of children's height were 70:<90cm.

Regarding distribution of the effect of the disease on studied children's general health, the current study revealed that the disease affected on less than half of children's general health such as; Delayed growth of weight and length, Behavioral problems, delayed social and intellectual skills, Skin related disease, Convulsions, and Bad odor of the body. Majority of children had not complication of the disease. All of children were made Phenylalanine level in the blood as regular investigation. Half of children made Phenylalanine level in the blood once every two months this result is may be due to this disease effect on normal development of children in their mental and their bodies.

Also this result disagree with the study done by **Tiele et al. (2019)** who studied "Investigation of pediatric PKU breath malodor, comparing glycol macro peptide with phenylalanine free L-amino acid supplements and they found the majority of children's general health such as; Convulsions, Delayed growth of weight and length, delayed social and intellectual skills, as well Bad odor of the body.

Also, this finding result is in agreement with the study done by **Walkowiak et al. (2019)** who studied "General health in classical phenylketonuria children": A retrospective cohort study. Advances in medical sciences and they found that the majority of children were measure Phenylalanine level in the blood regularly. Two third of the children assess their Phenylalanine level in the blood once every two months.

Regarding distribution of the studied children regarding their nutritional lifestyle, the current study revealed that more than half of the children were taking solid food after one year. Two third of children were taking three meals per day and the largest meal is the lunch. Nearly half of the children preferred food were Carbohydrates (rice – bread). While Majority of the children were taking the last meal from 6pm to 10pm. Three quarter of the children were receiving adequate fluids and the majority were receiving Permitted fluids (water – low sugar fruits). This result may be due to that during the first year of child age affected by disease the family become afraid from the food containing PHE so the mother give artificial food after one year only to keep the child health, also the child take three meals per day and the largest one was lunch because the child take the formula only in the morning so they become hungry and take the largest diet at the lunch time and also the lunch meal contain the preferable food for the children that rich with carbohydrates.

This result is in agreement with the study done by **Demirdas et al. (2017)** who studied "Micronutrients, essential fatty acids and bone health in phenylketonuria. Annals of Nutrition and Metabolism" where they found that one third of children were taking three meals

per day and the largest meal was the lunch. Majority of preferred food for children were Carbohydrates (rice – bead) and half of children were taking solid food after one year.

This result is in agreement with the study done by **Tonon et al. (2019)** who studied "Food Neophobia in Patients With Phenylketonuria" and found that two third of the children were Receiving adequate fluids and the majority of children were receiving the Permitted fluids (water – low sugar fruits).

Regarding distribution of the studied children towards their prescribed diet system, the current study revealed that the majority of children had followed special diet system. All of mothers prepared food for their children. Three quarter of children were taken diet system Free from protein. Two third of the children were Complied with the diet system. Majority of children were dispensing the prescribed diet once every month. Majority of children were dispensed amount of the prescribed diet. Majority of children's mothers were using calculation method to measure amount of foods, and nearly half of the children's mothers calculate amount of foods from the metabolic clinic. Majority of mothers were keeping the prescribed diet out of refrigerator. This result may be due to that the mothers were interested by diet system for their children.

This result is agreement with the study done by **Medford, Hare & Wittkowski, (2017)** who studied "Diet system influences on treatment adherence for children with PKU": a systematic review and found that majority of children were Complied with the diet system, and majority of children were dispensed the amount of the prescribed diet.

This result in agreement with the study done by **Singh et al. (2016)** who studied "Updated, web-based nutrition management guideline for PKU": an evidence and consensus based approach as they found that three quarter of children were Using calculation method to measure amount of foods, and all of mothers were keeping the prescribed diet out of refrigerator.

Regarding distribution of the studied children according to their types of education, the current study revealed education states that about three quarter of children not enrolled to nursery and less than half of them reported the main cause was late speech development for their children. Also, Minority of them didn't able to cope due to fear from others in the nursery. The main cause of refusing taking foods in the nursery was due to Unawareness of the teacher to the importance of eating the prescribed foods for such group of children. This result may be due to that the child take food from other children in the nursery and harm their self's and also unawareness of the teachers about the diet of the child, also among thous children due to the late speech development.

This result is in agreement with the study done by **Okano et al. (2016)** who studied "Education status of patients with phenylketonuria in Japan" and they found that majority of children not enrolled to nursery and nearly half of them reported that the main cause was late speech development for their children. This result is in agreement with the study done by **Lam & Davis, (2017)** who studied "Inborn Metabolic Disorders and Endocrine Disorders" and found that majority of children enrolled to nursery to be able to cope with others in the nursery.

Regarding correlation between compliance with the prescribed diet and

effect general health, the current study revealed that there is a highly statistically significant correlation between compliance with the prescribed diet and effect on general health. This result may be due that the child take the diet ratio of PHE is normal so not affected the health status of the child. This result is in agreement with the study done by **Medford, Hare & Wittkowski, (2017)** who studied "Diet system influences on treatment adherence for children with PKU": a systematic review and they found that there is a highly statistically significant correlation between compliance with the prescribed diet and effect of the disease on the children general health.

This result in agreement with the study done by **Jani et al. (2017)** who studied "Protein intake and physical activity are associated with body composition in individuals with phenylalanine hydroxylase deficiency. Molecular genetics and metabolism" and they found that there is statistically significant correlation between compliance with the prescribed diet and the effect of the disease on the children general health.

Conclusion

From the result of present study, it can be concluded that phenylketonuria had a negative effect on the children's lifestyle especially growth and development and general health, while had slight effects on sleep pattern of children.

Recommendations

In the light of the results of this study, the following recommendations were suggested:

- Emphasis the importance of regular follow up for mothers of children with Phenylketonuria for regular assessment and meet their actual needs to improve their children lives.

- Encourage children to be enroll in a nursery to increase their self-confidence through mass media as television, newspaper, posters, booklets and other communication channels.

- Encourage behavioral adjustment of the children suffering from Phenylketonuria through, decrease stressing events for children to avoid nervous behavior, make children's diet more attractive to encourage them to eat, improve home environment that support good behavior and increase teacher's awareness about diet and behavior of children.

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