

Comparison of face to face and non-face training methods by parents on the children serum phenylalanine levels with Phenylketonuria

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Abstract

Introduction: Phenylketonuria (PKU) is a congenital metabolic disorder of Phenylalanine, If which, the diet is not to be followed exactly, it can lead to progressive mental retardation. The purpose of this study was to investigate the effect of face to face and non-face to face trainings by parental on controlling serum phenylalanine levels in the children with Phenylketonuria.

Materials & Methods: In the clinical trial, 45 patients (aged 1-18 years) with Phenylketonuria, having the inclusion criteria, along with their parents, were divided into three groups of 15 subjects randomly; intervention 1: face to face training, intervention 2: non-face to face training using booklets and pamphlets and 3: a control group. The knowledge of parents was assessed and recorded by a questionnaire; and phenylalanine levels in children were examined by a venous blood samples before and after intervention in all households. The paired t-test, Chi-square and ANOVA and Tukey tests were used for analyzing data.

Results: There was no significant difference for parents' knowledge prior to the intervention in the groups, but after the intervention, there was a significant difference in the intervention group compared to the control group ($p = 0.006$). The educational intervention (face to face and non-face to face trainings) enhanced the knowledge of the family. This increase was seen in the control group ($p = 0.086$). For serum phenylalanine, although both educational methods led to reduce, but this reduction was significant only in the non-face to face method ($p = 0.015$).

Conclusion: This study showed positive effects of both educational methods on the amount of parental awareness; however, the non-face to face educational effectiveness of parents on phenylalanine levels of children was showed more outcomes. Therefore, the regular non-face to face education parameter should be included to follow-up of these patients.

Keywords: Dietary intervention, Face to face and non-face to face training, Phenylalanine, Phenylketonuria (PKU).

Introduction

Phenylketonuria (PKU) is one of the most common metabolic diseases caused by deficiency of the enzyme phenylalanine hydroxylase, which is created as a result of disorder in the amino acid phenylalanine metabolism and can lead to progressive mental retardation (1). The gene responsible for this disorder was found on chromosome 12. The patients are normal at birth. However, if patients are not properly diagnosed and treated, gradually their evolving abilities are impaired, and consequently they will have severe mental retardation, which it may not be

so clear in the first months of life (5). Bichel et al proposed the use of diet for the treatment of this disease. He found that with the exception of mental retardation, the rest of the clinical problems of patients treated are reversible by restricting phenylalanine in the diet (6). In some developed countries, more than 50 years have passed since the screening of the disease (7,8). Screening for the disease started recently in Iran for infants, which it poses the need to inform public about the disease.

The treatment of this disease is on the basis of the limitations for taking phenylalanine in the diet that is possible using semi-synthetic

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products. With the maintenance of serum phenylalanine levels in the range of 2-6 mg/dl-recommended for children under 12 years and 2-10 mg/dL for children above 12 years old (9,8), the disease control will be satisfactory. Much attention on food limitation causes to receive insufficient food so that parents are regularly concerned for the permanent starvation of their child. The reasons for this deficiency in Iran are: the lack of parental awareness; the lack of food for the patients; and finally being the expensive price of these products (7). The best outcome of health for children with special needs is related to the family functioning (8). The parents who are aware of children's special needs, they will make greater efforts to meet these requirements (10). Since patients with PKU and chronic diseases are largely young population, the role of parents in their care is of great importance (10).

Before creating a dangerous condition and irreparable problems from the disease, education for Parents to avoid of any difficulty, or can lower its severity to a minimum. Given that children physically and psychologically are not able to take care of themselves, therefore, the parents are the individuals who have to be aware of children's needs and principles of caring them. Because of professional responsibility, nurses are the most appropriate people for educating the parents and supporting the family (11).

A training with the aim of acquiring knowledge and skills leads to an increase in the ability of individual health decisions and behavior change (12,13). In a categorization, education has been divided into two types of face to face and non-face to face categories.

One method of face to face training is lecture scheme that is economically advantageous. One of the methods of non-face to face training is using educational pamphlets and booklets that are non-face to face affordable and accessible, however it is not adequate for people who are illiterate (13). In reviewing previous studies on the effectiveness of training, teaching can be divided into three categories, 1) - a group of studies in which education has been found effective, for example, Naderi, (14). 2) - Some studies have found that such training is ineffective for example, Salmani-Dastjerd, (15). 3) - a group of studies suggest that both face to face and non-face to face trainings are the effects, and none has superiority like Maverdi's study (16). In the United States, a study has shown the effect of education for parents of patients with PKU on controlling and reducing the amount of serum phenylalanine in children with PKU (17). The studies conducted in the field of education and awareness of the parent of these patients and comparing the efficiency of these methods are limited, especially such studies have not previously been carried out in Iran. Therefore, this study has been conducted to survey and compare the effect of two educational methods of parents on serum phenylalanine level control of children with PKU.

Materials and Methods

In a randomized clinical trial in 2012, 45 out of 65 patients with PKU 1-18 years followed up in the reference clinic of PKU of the Khuzestan province, located in the Children's Hospital of Abuzar. The rest of the patients were excluded due to lack of regular visiting

and follow-up. A duplicated questionnaire containing essential information for parents about the care and needs of these patients, especially in resource-based regimen consisting of 19 questions of four-choice were developed.

The content validity method was applied to assess the scientific validity of questionnaire; first, considering the variables of research and published books and articles, the initial questionnaire was set up, and was shown to 10 teachers from Ahvaz Faculty of Nursing and Midwifery; then, corrective feedback was collected and after the necessary amendments, the final questionnaire was developed.

The test re-test method was applied in order to determine the reliability of the questionnaire. In this study, 20 parents randomly were selected; the questionnaire was distributed, completed, and collected from this group and again after two weeks, the questionnaire was given to the same people in which the reliability with a coefficient of Cronbach's Alpha was 90%. After ensuring to be literate all the subjects, and to describe how to respond, questionnaires were given to the parents and were completed at the place of the clinic. Parents were randomly divided into three groups, 1) - intervention by direct education 2) - and 3) - intervention indirect educations - control groups (A total of 45 subjects; 15 patients in each group). For measuring the serum phenylalanine of all children, the venous sampling was performed at a reference laboratory in the Abuzar Children's Hospital. Before the intervention, two milliliters of fasting venous blood were taken in the morning, and then, five drops of blood were placed on the Guthrie card filter paper (produced by Kimia Co.) with sensitive

0.5mg/dL. Twenty-four hours after drying using the boiling temperature machine, Equipments, (Azma Gostar Co.), the droplets are heated for one hour at 110 degrees; then for an hour, they are carried to the shocking device, Stat Fax (Aware Enss Co.); finally serum phenylalanine levels were measured and recorded by an expert using El & 800 (Biotech Co.). Acceptable levels of phenylalanine in children under 12 years of age and over 12 years are considered as 2-6 mg/dL and 2-10 mg/dL, respectively (8). In the intervention Group 1, parents were under training, with over eight meetings of lecture and question and answer of two-hour weekly. The intervention Group 2 was under training a weekly basis for one month with the use of educational pamphlets, handouts.

The Group 3 (control) was followed- up only with the previous usual cares and follow-ups and without any additional training intervention. In order to measure the sustainability of intervention, one month after the last face to face training session and one month after the delivery of educational materials, parents were asked again to answer questions and return back the questionnaires. At the same time for serum phenylalanine after intervention, the venous blood sampling of children was performed again. Information for patients and parents, including date of birth of patients and parents, age at diagnosis, the parents' education level (a division of the below diploma, high school diploma, and above diploma), level of family monthly income, location of family life were collected.

The statistical analyzes were performed using SPSS 19. The numbers were presented as Mean \pm SD. The quantitative variables were compared using "analysis of variance"; and for

qualitative variables using "chi-square". A pairwise comparison of the two groups was done using 'Tukey test'; variables before and after intervention were compared using the Paired t-test. p-values of less than 0.05 are considered as significant.

After explaining the purpose and method of the study, a written consent was obtained from all the parents for entering the study without publishing personal information. This study did not impose additional sampling or referring to the patients and families to make a dent in the treatment program. This study was confirmed at the Research Ethics Committee of Ahvaz Jundishapur University of Medical Sciences.

Results

Of the 45 patients studied, 28 (62.2%) and 17 (37.8%) were male and female, respectively. The mean age of patients was 7.84 ± 4.76 years. 60% of patients (27 patients) had familial married parents. In terms of age at the time of diagnosis at time of birth, less than six months, more than six months were three cases (6.7%), seven cases (15.7%), and 35 cases (77.8%), respectively. Twenty persons (44.4%) Of the patients were living in the Ahvaz County and 25 people (55.6%) were residing in other Counties.

The intervention and control groups were compared in terms of effective variables that represent groups had the same condition before intervention and proper distribution of patients in the various groups.

The achieved p values were obtained in terms of age, the mother's age, $p=0.305$, the father's age, $p=0.061$; the mother's education, $p=0.167$, the father's education, $p=0.086$;

monthly parental income, $p=0.089$; previous parental education, $p=0.245$; the child's age at the diagnosis time, $p=0.673$; and location of life, $p=0.914$.

Before the intervention, the average score of the parents' test, and serum phenylalanine levels of the patients were 8.46 ± 5.36 , and 9.26 ± 6.48 , respectively. The amount of parental test score prior to the intervention in the various groups did not have significant differences ($p=0.817$); furthermore, in terms of the serum phenylalanine, groups were the same ($p=0.065$).

After the intervention, in the trained groups, the parents' knowledge increased, and phenylalanine levels decreased (Tables 1 and 2). With only two thirds of patients and parents taught, it improves the clarity of variable individuals.

Table 3 shows details of the main variables before and after the intervention of the subjects in the all cases. Although only two-thirds of the patients and their parents were trained, it makes a clear improvement in the variables of the all cases. On the information after the intervention, control group (despite the lack of intervention in this group) are also included. However, change is clearly visible with the intervention.

In a pairwise comparison of groups, in terms of the amount of parental awareness after the intervention, only the face to face training Compare with the control group showed a significant difference ($p = 0.04$). This is a comparison about the amount of serum phenylalanine after intervention which was different between the two groups of face to face training and non-face to face training ($p = 0.023$).

Table 1: Comparison of the amount of parental knowledge of patients with PKU according to the test, before and after the educational intervention in the intervention and control groups

| Treatment group | Intervention1 Mean±SD | Intervention2 Mean±SD | Control Mean±SD | p value |
|--|--------------------------|--------------------------|--------------------|---------|
| Parental knowledge before intervention | 9.06 ± 5.70 | 8.53 ± 5.22 | 7.80 ± 5.45 | 0.817 |
| Parental knowledge after intervention | 14.06 ± 3.78 | 11.60 ± 4.51 | 8.46 ± 5.20 | 0.006 |
| p value | <0.001 | <0.001 | 0.086 | |

Table 2: The serum phenylalanine levels in patients, before and after the educational intervention in the intervention and control groups

| Treatment group | Intervention1 Mean±SD | Intervention2 Mean±SD | Control Mean±SD | p value |
|--|--------------------------|--------------------------|--------------------|---------|
| Phenylalanine before intervention(mg/dL) | 6.87 ± 4.19 | 11.50 ± 6.10 | 9.42 ± 5.30 | 0.065 |
| Phenylalanine after intervention(mg/dL) | 5.88 ± 3.43 | 9.73 ± 4.68 | 7.93 ± 3.20 | 0.03 |
| p value | 0.247 | 0.015 | 0.43 | |

Table 3: The test score status and serum phenylalanine levels of all contributors in the study before and after training in the parents and children with PKU

| | Test score* | | | The mean score | Serum PKU | | The mean serum(mg/dL) |
|----------------------|-------------|-------|-------|----------------|------------|----------------|-----------------------|
| | >15 | 10-15 | <10 | | Controlled | ‡Inappropriate | |
| Before intervention | 2.2% | 46.7% | 51.1% | 8.46 ± 5.36 | 37.8% | 62.2% | 9.26 ± 5.48 |
| After intervention § | 20% | 48.9% | 28.9% | 11.37 ± 5 | 46.7% | 53.3% | 7.85 ± 4.06 |

*Of 19 scores of total test, ‡ it is applicable for serum level higher than 6 mg/deal in the age under 12 years and above 10 mg/dL above 12 years

Discussion

The Parental education caused a significant increase in the parental knowledge, but improvement of serum phenylalanine which is the most important criterion to control these patients was diminishing; however, this change was less than the expected limit.

The most important health point for maintaining health of patients with PKU is observance of food regimen in the restriction of phenylalanine. Although usually at the beginning of the disease diagnosis, important tips in caring for these patients given to the family but like other chronic diseases,

especially diseases that are associated with the patient's lifetime, the disease control and long-term care are costly and overwhelming. Except for food needs (milk, and Phenylalanine Free food products), pharmaceutical, Periodic testing, physiotherapy, occupational therapy, speech therapy, which are the basic needs of these patients, cases such as social support, education and special education ... should be considered. The involvement of family and patient in the mass of requirements mentioned sometimes causes to forget important nuances in the treatment and inappropriate control of these patients. The periodical training for parents (and possibly for children) by default should be able to cause clear improvement in measures of disease control such as phenylalanine level in these patients.

Hamidzadeh is believed that intervention of training for parents and other family members involved in the care of the child is important (18). Mac Donald et al. United States, 2008 showed the positive impact of education on phenylalanine levels pediatric patients (17). The positive impact of education on the chronic disease process (such as thalassemia and asthma) is rational (19, 20). Mc Naull et al. recommended that the combination of both educational methods is an effective way of learning for parents (21).

Wasbran et al., by an analysis of 40 studies, showed that the elevated serum phenylalanine levels during treatment, especially in the first 12 years of life cause to reducing a significant loss in IQ (22). The study of Potensik et al. in Australia showed that those patients who adhere to their diet entirely up to age 10 are less likely to have the neural defects. However, even in the best circumstances,

these patients when entering the school have an IQ lower than their healthy sisters and brothers (23). This study shows that the Khuzestan Province (Southern Iran), the majority of these patients (60%) are due to consanguineous marriages; owing to the autosomal recessive inheritance of the disease, the rate of the disease occurrence can be decreased by preventing consanguineous marriages.

In the study, the diagnosis age of this disease was in the majority of patients over six months that has remained significant degrees of irreversible mental retardation. In the study of Maylan, the diagnosis age of patients by pediatricians was from one month to 22 years that at the time of diagnosis, 94.5% of patients were with mental retardation and 48.9% had epilepsy (24). The start of the PKU screening program in the Iran that during the study has been begun in the Ahvaz, and is expanding, will prevent this disaster.

The Phenylalanine level control is a reflection of proper observation of nutritional tips in restricting the phenylalanine and preparation of essential dietary needs. Although, this seems simple, but it is a function of many confounding factors such as social status, the residence place, accessing to the health services, education, the family support, coverage of state and insurance health services, and finally effort of parents and other family members in the accompaniment and periodical reference, the necessary tests, and to follow medical and nutritional instructions of the PKU team. The periodical training of the family as the default should be effective to improve many of these factors. The present study showed that, parents before education answered on average to less than half of the

test questions. After learning, the response rate was much better; this study showed the need for training in this population. The rate of the Phenylalanine serum in the studied children was sufficient in only 37% of cases that after education (only two-thirds of the parents, Table 3); the rate has increased to nearly 47%.

In this study, for the high percentage of the rural population and away from the center, being a high population of households, economic status lower than in larger cities, many problems in achieving a low-protein meal and replacements of protein cause more difficult control of disease in this population. Therefore, in such circumstances, education and psychological- social support will be more essential

Bekhof et al. in surveying the effect of awareness on diet satisfaction has considered the role of psychological and emotional factors complex and complicity; furthermore, the study has considered phenylalanine serum control as a very important factor (25). Parents of involved children have an important role in helping the child for understanding and responsibility of their diet. The most families took part in this study were not well educated, thus understanding the replacement system of phenylalanine and use of it is difficult and most of them cannot calculate the amount of phenylalanine that children intake. Parent's training and increase of their awareness and continuing educational programs ensure authors that patients and their families have understood the concept of diet. In addition, findings of MacDonald et al. have identified that serum phenylalanine level will be in the normal range following an increase of parents' awareness (17). Also in the study of Ozel et al., children's serum phenylalanine was higher

for whom mothers had fewer score in awareness evaluation than mothers who had higher scores (26).

Hamidzadeh in a study on the effect of mothers' awareness of anthropometric changes in the infants showed that the mean difference of awareness in both ways of face to face and non-face to face education was not significant but when face to face is composed with a non-face to face way will be effective (18). In this research, non-face to face education was more effective in reducing serum phenylalanine. This difference may be due to more durable and repeatable non-face to face than verbal education. Yet other people except parents can benefit of this education that be more effective indirectly in the more control of phenylalanine serum level and disease. The number of patients in each group was one of the limitations for this study; although, the families did not leave the study. It should be noted that this disease is completely rare; however it is more abundant in the Khuzestan and similar regions only due to the high percentage of family marriage. It is impossible to include patients who are hundreds of kilometers away from the center, and some of them are living in other provinces in these kinds of studies. In spite of this number seems low, but according to the total number of patients followed up in the country, it is considered as a significant number. In these such cases that the disease prevalence is low, multi-center studies certainly will give better results. Another limitation of this study was the possibility of communication and exchange of information between the groups that could affect in the evaluation of the final result of training in different groups. Multi-center studies and more research are needed

for evaluating the impact of training and preparation of recorded educational programs to different segments of society, especially for the parents.

Conclusion

Generally, parental education in any case, caused to improve household data of patients with PKU and ultimately better control of the disease. Contrary to popular belief, despite a lack of active training in non-face to face training, its effectiveness in the control of serum phenylalanine level was more than face to face training. Similar studies with the presence of more patients and families may produce different results. Finally, with regard to the non-face to face training, which is

cheaper and is equally available for the whole family in far and near cities, this action should be included periodically in the care plan of patients with PKU, especially in patients poorly controlled.

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