

Effects of A Phenylketonuria Camp on Knowledge and Attitudes of Adolescents with Phenylketonuria

Fenilketonüri Kampının Fenilketonüri Adölesanların Bilgi ve Davranışları Üzerindeki Etkileri

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ABSTRACT

Objective: This study aimed to determine the effect of a phenylketonuria camp on adolescents with phenylketonuria (PKU).

Material and Methods: A small cohort-type study was carried out with 67 participants between the ages of 12-16. Participants were selected from PKU patients who were attended a PKU camp, organized for patients with phenylketonuria, aiming socialization, disease-related information transfer, and providing pleasant time with various social activities. Besides the patients, specialists of pediatric metabolism, psychologist and dieticians attended the camp. The study questionnaire was applied by a face-to-face interview on the first and last days of the camp. The primary outcome Questions were asked about PKU perception, and knowledge scores were calculated for each participant by adding the number of correct answers in the 10-item test.

Results: Of the participants, 34 (50.7%) were females, while 33 (49.3%) were males. Twenty-one (31.3%) patients had another family member with phenylketonuria. The mean knowledge scores were already high at the beginning of the PKU camp (median 8, min. 4, max. 10). When the patients' knowledge of PKU was compared, there was no significant difference between before (7.40±1.62) and after (7.72±1.80) the camp (p=0.097). However, it was found that the opinions of the participants about the difficulty of being a PKU patient improved significantly (p= 0.039).

Conclusion: A PKU camp for adolescents is effective in improving the perceptions of the participants. However, it would be beneficial to increase the duration and frequency of PKU camps to achieve better outcomes.

Key Words: Adolescents, Phenylketonuria, Socializing, Education of Patients, Phenylketonuria Camp

ÖZ

Amaç: Bu çalışmada fenilketonüri kampının fenilketonüri (PKU) adölesanlar üzerindeki etkisinin değerlendirilmesi amaçlanmıştır.

Gereç ve Yöntemler: Bu kohort tipi çalışma, yaşları 12-16 yaş arasında değişen toplam 67 katılımcı ile gerçekleştirildi. Katılımcılar, fenilketonüri hastalar için düzenlenen, hastaların sosyalleşmesini, hastalıkla ilgili bilgi düzeylerini arttırmayı ve çeşitli sosyal aktivitelerle keyifli vakit geçirmeyi amaçlayan PKU kampına katılan PKU hastalarından seçildi. Kampa hastaların yanı sıra, çocuk metabolizma uzmanları, psikolog ve diyetisyenler katıldı. Araştırma anketi kampın ilk ve son günlerinde yüz yüze görüşme ile uygulandı ve 10 adet bilgi soruları sorularak, her katılımcı için doğru cevap sayısına göre bilgi puanı hesaplandı.



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Bulgular: Katılımcıların 34'ü (% 50.7) kız, 33'ü (%49.3) erkekti. Yirmi bir (%31.3) hastada fenilketonüri hastası başka bir aile üyesi vardı. Ortalama bilgi puanları PKU kampının başlangıcında da yüksek (ortanca 8, en az 4, en fazla 10 puan) olup, hastaların PKU bilgisi karşılaştırıldığında, kamp öncesi (7.40 ± 1.62) ve sonrasında (7.72 ± 1.80) arasında anlamlı bir fark bulunamadı ($p= 0.097$). Ancak katılımcıların PKU hastası olmanın zorluğuna ilişkin görüşlerinde kamp sonrası anlamlı bir iyileşme olduğu görüldü ($p= 0.039$).

Sonuç: Ergenler için düzenlenen PKU kampları, PKU hastalarının algılarını iyileştirmede etkilidir. Ancak, daha iyi sonuçlar elde etmek için PKU kamplarının süresini ve sıklığının arttırması faydalı olacaktır.

Anahtar Sözcükler: Adölesanlar, Fenilketonüri, Sosyalleşme, Hastaların Eğitimi, Fenilketonüri Kampı

INTRODUCTION

Phenylketonuria (PKU) is a rare metabolic disease with autosomal recessive transition characterized by the failure of phenylalanine-tyrosine conversion due to the lack of phenylalanine hydroxylase, its accumulation in blood, resulting in central nervous system findings (1,2).

If babies with this disease are not timely diagnosed and treated, some physiological, neurological, and intellectual disabilities arise, including irreversible brain damage, mental retardation, developmental delay, seizures, involuntary contractions, hyperactivity, motor deficits, gait disorders, and autistic behaviors (2). If treatment is started in the first four weeks of life, people with PKU can usually live as independent adults (3).

The prevalence rates of PKU vary between countries. Figures of 1/15.000 and 1/10.000 were reported from the United States and Europe, respectively (2,4). Turkey has a relatively high prevalence of PKU, reaching 1/4.000 (5).

Success in PKU treatment is directly correlated with keeping the blood phenylalanine (Phe) levels low, and it depends on patients' and their parents' ability and motivation on lifestyle changes, and the adherence to the low phenylalanine diet (6,7). Severe emotional disturbances and social withdrawal have

been reported in adolescents and young adults with PKU who have resumed a regular diet (8).

A PKU camp has been organized regularly since 2014 in Turkey, in which patients with phenylketonuria come together, receive training about their illness, and have a pleasant time with various social activities.

This study aimed to evaluate the effectiveness of a PKU-camp and to measure the change in the knowledge and attitudes of the participants before and after the camp.

MATERIAL and METHODS

A small cohort study was performed after obtaining ethical approval (numbered: GO 18/799-20) from the Hacettepe University Faculty of Medicine Clinical Research Ethics Committee. The study was carried out in Hacettepe University Faculty of Medicine, Department of Pediatric metabolism.

The PKU camp is traditionally organized by METVAK (<https://www.metvak.com/>) since the year 2014. The fifth camp was held between 5-11 September in 2018 in Uludağ (Turkey) to improve the social and physical skills and knowledge of children with PKU. Besides the patients, specialists of pediatric metabolism, psychologist and dieticians attended the camp.



Figure 1-2: Different activities in PKU camp.

With the guidance of dieticians, PKU patients learned to prepare most of their favorite tastes fitting their diet. Also, they had fun with different activities (Figure 1,2). Five PKU children attending previous camps guided their friends.

The study participants consisted of patients between the ages of 12-16 who were followed up with the diagnosis of classical phenylketonuria in Hacettepe University Department of Pediatric Metabolism and attended the PKU Uludağ Camp in 2018. All 67 participants of the camp were invited to join without sampling.

The primary outcome variable of the study was the “perception of comfort with PKU.” Independent variables were age, sex, number of people living at home, number of siblings, school grade, the presence of another PKU patient in the family, and knowledge and behaviors concerning PKU. Data collection was done in a convenient room using a questionnaire applied by a face-to-face interview on the first and last days of the camp. The Cronbach’s Alpha internal consistency value of the knowledge questions (10 items) in the post-test was 0.596. Mean knowledge scores were calculated by adding the number of correct answers.

Statistical Analysis

The Statistical Package for the Social Sciences software (SPSS for Windows, Version 25.0, Chicago, IC, USA) was used for statistical analysis. The results were presented as frequency and percentage for categorical variables and mean and standard deviation for numerical variables. The suitability of numerical variables to normal distribution was evaluated using the Kolmogorov-Smirnov test.

Numerical variables were compared with the Wilcoxon Signed Ranks test. Besides, the Marginal homogeneity or McNemar tests were used for comparing dependent categorical variables. A p-value of <0.05 was considered sufficient for statistical significance.

RESULT

Thirty-four participants (50.7%) were female, while thirty-three (49.3%) were male. Twenty-one (31.3%) patients had another family member with phenylketonuria. Most of the patients (80.6%) had siblings. Likewise, most (n= 45, 67.2%) lived in nuclear families.

The mean knowledge scores were already high at the beginning of the PKU camp (median 8, min. 4, max. 10). When the patients’ knowledge of PKU was compared, there was no statistically significant difference between scores before (7.40 ± 1.62) and after (7.72 ± 1.80) attending the camp ($Z=1.658$, $p=0.097$).

It was found that the opinions of the participants about the difficulty of being a PKU patient changed significantly at the end of the camp (Table I). However, there was no statistically significant change in mood (Table II).

When the participants’ perception of social support and their views on the camp were compared before and after the camp, there was no statistically significant difference (Table III).

DISCUSSION

At the end of the PKU camp, the participants’ views on the difficulty of being a PKU patient changed positively. The knowledge scores of the participants on PKU were already high at the beginning of the camp.

Neurocognitive and psychosocial impairment can occur even if the treatment of PKU is initiated on time (9,10). In a meta-analysis, Moyle et al.(11) showed that patients with PKU were significantly different from controls concerning full-scale IQ, processing speed, attention, inhibition, and motor control. In addition, psychological disorders such as low self-esteem, low motivation for success, decreased autonomy, and decreased social competence have been reported in these children

Table I: Comparison of perception of comfort in PKU patients.

	Slightly difficult n (%)	Moderately difficult n (%)	Highly difficult n (%)	MH*	p
Before camp	18 (32.4)	17 (30.4)	21 (37.5)	2.064	0.039
After camp	24 (42.1)	19 (33.3)	14 (24.6)	-	-

*Marginal Homogeneity test

Table II: Comparison of the answers to the question of “How do you feel in general before and after the camp?”

	Desperate n (%)	Unhappy n (%)	Cheerful n (%)	Very cheerful n (%)	Undecided n (%)	MH*	p
How do you feel in general? (Before)	3 (4.5)	9 (13.4)	29 (43.3)	16 (23.9)	10 (14.9)	0.717	0.473
How do you feel in general? (After)	3 (4.5)	7 (10.4)	35 (52.2)	16 (23.9)	6 (9.0)	-	-

*Marginal Homogeneity test

Table III: Comparison of participants' views on the PKU camp.

	No n (%)	Yes n (%)	Not sure n (%)	MH*	p
Having phenylketonuria is not a problem for me				0.503	0.615
Before	12 (17.9)	42 (62.7)	13 (19.4)		
After	15 (22.4)	39 (58.2)	12 (17.9)		
My family supports me for my illness				0.378	0.705
Before	2 (3)	60 (89.6)	5 (7.5)		
After	0	63 (94)	4 (6)		
It does not bother me that my friends know my illness				0.870	0.384
Before	7 (10.4)	47 (70.1)	13 (19.4)		
After	13 (19.4)	40 (59.7)	14 (20.9)		
Will it be good for you to be with your friends who have the same disease?				0.277	0.782
Before	5 (7.5)	59 (88.1)	3 (4.5)		
After	5 (7.5)	60 (89.6)	2 (3)		
Do you think the camp will be useful to you?				1.069	0.285
Before	3 (4.5)	60 (89.6)	4 (6)		
After	2 (3)	58 (86.6)	7 (10.4)		
Would you like to rejoin the camp?				1.265	0.206
Before	-	56 (83.6)	11 (16.4)		
After	-	60 (89.6)	7 (10.4)		
Will you meet your friends after camp?				0.180	0.857
Before	7 (10.4)	40 (59.7)	20 (29.9)		
After	3 (4.5)	47 (70.1)	17 (25.4)		
Would you like to work in the phenylketonuria camp in the future?				0.365	0.715
Before	7 (10.4)	46 (68.7)	14 (20.9)		
After	3 (4.5)	52 (77.6)	12 (17.9)		

*Marginal Homogeneity test

(12,13). Attention was drawn to the risk of depressive mood, general anxiety, and social isolation in adolescents and adults (9,12,13).

It has been stated that, in the treatment of phenylketonuria, going beyond focusing on metabolic control, adopting a holistic approach that supports the psychosocial wellbeing and quality of life of the child and family can help achieve more productive and successful results (14). Healthcare camps, which can be considered as part of the holistic approach, are thought to be beneficial because PKU reduces the quality of life of the patient and his/her family (15).

Health camps have been reported to be useful for children with cancer and advantageous regarding many psychosocial health indicators (16). It has been argued that these benefits of camps may also apply to children with similar chronic conditions (16).

Health camps are effective in patients with Type 1 diabetes mellitus (DM), a disease requiring dietary regulation, similar to PKU (17–19). Camps have been organized where children and adolescents with Type 1 DM, a relatively rare disease, can meet other akin patients and receive training on their diet and treatment to achieve positive improvements in their health indicators (17). According to the American Diabetes Association (ADA), more than 30.000 young people in North America and about 16.000 in other countries attended these camps, whose content and size have evolved over time (20).

In a study in Korea, aiming to determine the effect of the health camp program on juvenile diabetes mellitus patients on self-efficacy, patient-role behavioral adjustment, and glucose metabolism, the health camp has been reported to increase self-efficacy and patient-role behavioral adjustment scores in juvenile diabetes mellitus patients, except for glucose metabolism (18).

In another study investigating the effectiveness of the diabetes camp, Semiz et al. reported that the health camp increased the knowledge of the participants and the ability to manage the disease (19). Furthermore, it was observed that the improvement in the nutritional and diabetic knowledge of children attending consecutive camps was more pronounced in the second camp compared to the first camp. However, there was no significant change in HbA1c levels.

It has been mentioned that, the nutrition health camp for obese children positively affects nutritional attitudes (21). Besides, other researchers stated that the health camp was moderately useful in solving emotional and behavioral problems in children (22).

In a study evaluating asthma camps with the participation of 174 participants, the level of knowledge and self-management was high at the beginning. However, a further significant increase was detected at the end of the camping period (23).

The PKU camp is considered a suitable environment to introduce patients to similar peers. In this way, the person may realize that he/she is not alone and can take advantage of the experiences of other patients. The researches summarized above supports the idea that the PKU camp should provide favorable results for adolescents. Nevertheless, it was surprising that the significant difference in the results of our survey was detected only in the perception of the disease difficulty. However, even this achievement can be considered a success.

Nonetheless, we consider that more studies are needed to reveal the underlying causes of the camp's inability to make a significant difference in other matters and increase its effectiveness. Hence, the content of the camp should perhaps be revised and improved with the feedback of the participants. It has been reported that when organizing camps for children with chronic diseases, understanding children's perspectives will help researchers and campers to increase the effectiveness of the camp (24). Future studies should check whether the questionnaire used is correctly perceived by patients with PKU, a potential intellectually disadvantaged group.

One reason for not making any difference in knowledge questions before and after the camp may be that the participants already have sufficient information about their diseases. Thus, we may claim that patients with PKU, who were followed up in our hospital, had a favorably good education about their conditions. Still, more studies are needed to clarify this unexpected finding. Another possibility is that the information in the camp could not be transferred from the dietician or trainer to the patient. To eliminate this probability, support can be obtained from educators who train special groups, such as children with PKU.

Limitations

The data collection tool of this study was prepared by the researchers and not tested before. Standardized and validated instruments such as the Phenylketonuria – Quality of Life (PKU-QOL) could provide more reliable results. Data collection was performed by a researcher non-familiar with the participants, which could have caused anxiety and hesitation in this vulnerable population. The intellectual capacities of the participants were not assessed. Since PKU has the potential to deteriorate cognitive functions, related limitations might be present.

CONCLUSION

The PKU camp held in Turkey had a positive impact on patient perceptions, but the effectiveness of the camp needs to be increased. Better study outcomes and effective results may be achieved if the contents of the camp are tailored according to the needs, expectations, knowledge, perceptions, attitudes, and skills of the participants. This study will make significant

contributions to the development of new scales to be used in future studies.

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