Parent knowledge regarding food selection for children with PKU: Results of a survey in the United States

Rodolfo Ramos-Álvarez[1,*], Maili Kapp [2], Amaya Bélanger-Quintana [3], María Mercedes Rodríguez-Ruiz [4], César Augusto Solano-Galvis [5], Antonio Campos Soto [6], Kirsten Ahring [7] and Susan E. Waisbren [8,*]

[1] Department of Social Psychology, Melilla Campus, University of Granada, 52005 Melilla, Spain
[2] Teaching Innovation Project (Code: 14–82), Melilla Campus, University of Granada, 52005 Melilla, Spain; mailikapp@correo.ugr.es
[3] Ramón y Cajal University Hospital, Spain; amaya.belanger@uah.es
[4] Osakidetza, Basque Health Service, 010006 Vitoria, Spain; rrmertxe@gmail.com
[5] Department of Experimental Psychology, Melilla Campus, University of Granada, 52005 Melilla, Spain; casolano@ugr.es
[6] Department of Didactics and School Organization, Melilla Campus, University of Granada, 52005 Melilla, Spain; antonio.campos@docentemelilla.educacion.gob.es
[7] Center for PKU, Copenhagen University Hospital, 2600 Copenhagen, Denmark; kirsten.ahring@regionh.dk
[8] Genetics and Metabolism Programs, Boston Children's Hospital, Harvard Medical School, Harvard University, Boston, MA 02115, USA
* Correspondence: rodolfo@ugr.es (R.R.-Á.); Susan.waisbren@childrens.harvard.edu (S.E.W.)

Abstract: Dietary treatment is the main therapy for most patients with phenylketonuria (PKU). Parental knowledge for food selection is crucial not only to ensure adequate metabolic control and brain development during childhood, but also to promote life-long dietary behavior in the offspring. The aims of this study were to assess the knowledge of parents or other caregivers of children with PKU regarding food selection in accordance with medical recommendations and to evaluate factors that influence their level of knowledge. An online or paper survey (n=178) was distributed to parents or caregivers of patients with PKU throughout the United States. It included a validated food selection questionnaire to assess if the respondent adequately identified foods that require certain restrictions vs foods that can be consumed freely by an individual with PKU. Results showed that the level of knowledge regarding the PKU diet was high with regards to forbidden or limited foods. Knowledge regarding allowed food was relatively low. Providing education regarding diet for PKU parents/ caregivers is important to ensure adherence and adequate metabolic control. It is important to include general concepts regarding the diet but also specifics regarding food selection, keeping in mind that some items might be more confusing than others. The results of our study can be used to improve diet education programs.

Keywords: phenylketonuria; PKU; diet; dietary knowledge; food selection

1. Introduction

Phenylketonuria (PKU) is a recessive autosomal disorder of phenylalanine (Phe) metabolism which, due to a partial or total inability to transform Phe into tyrosine, causes the accumulation of Phe in the body. High Phe concentrations have deleterious effects mainly in brain development and function [1]. PKU was the first inborn error of metabolism in which dietary therapy successfully prevented most of its symptoms [2]. In recent years, alternative treatments have become available, but up to now diet is still the main or only treatment for most individuals with PKU. Maintaining Phe levels within the recommended target range enables patients to have normal intellectual development and function. Individuals with PKU follow a diet that restricts any food containing toxic levels of Phe. These foods include all high protein containing products such as meat, poultry, fish, eggs, dairy, nuts, beans, most grains and many other foods [3]. Depending on each patient's individual tolerance to Phe, these restrictions are more or less severe. Other amino acids, calories or micronutrients are supplemented with special formulas.

For individuals with PKU, observing dietary rules is crucial. However, following the strict rules of this nutritional regimen remains problematic for many parents and patients. Poor adherence usually appears during adolescence and

worsens during adult life leading to poor metabolic control and neurological, psychological and social complications. It is acknowledged that despite the high efficacy of the dietary therapy, the outcome of patients remains suboptimal [4]. Moreover, physical complications may arise due to a poor nutritional status [5-6]. Poor adherence has multiple reasons that range from the bad taste of special products, difficulties in obtaining certain supplements, boredom with a very limited selection of foods or neurological difficulties in planning meals [7-10].

Following any diet, but especially one as demanding as a PKU diet, requires relevant knowledge on food composition, meal planning and specific nutritional recommendations [11]. Without this knowledge, patients and their parents cannot make informed decisions regarding their diet and health [12-14]. Which foods are selected to buy, prepare and eat is a central part of any dietary regimen. Food selection is determined by sensory aspects (taste, texture, appearance) but also age, gender, mood and very importantly by economical determinants, education and social environments [15-18]. These aspects affect food selection and neophobias also in PKU [19].

Dietary knowledge is initially acquired by parents and, subsequently, by their children who often carry on patterns established early on throughout adolescence and adulthood. The first years of life are therefore very important in shaping future conduct, and parents are the model for their children during these first experiences with food. Their attitudes will determine future behavior towards diet and have an impact on dietary adherence [19-20]. The mother's role seems to be especially relevant, as in most households she is the one that makes the nutritional decisions for her children [21]. Differences have been found between the way fathers and mothers implement alimentary practices and the way practices affect their children [22].

Educating parents/caregivers regarding the diet has been recognized by both families and physicians [23]. Some studies examine the impact of different socioeconomic and psychosocial factors on level of adherence to dietary recommendations [24-25]. Only a few publications investigated the impact of parental knowledge on diet adherence [26-27,13,19].

To our knowledge, this is the first study that analyzes parental/caregiver food selection for children with PKU.

2. Materials and Methods

2.1. Participants

Caregivers of children 0-18 years of age diagnosed with PKU living in the United States were asked to participate in this study. A food selection survey was distributed online among the members of the PKU Association of California and the National PKU Alliance (NPKUA). 13.6% of the answers were also obtained with a paper format of the same survey available at two patient meetings. One was organized by the California Coalition for PKU and Allied Disorders and the other was held in Oregon and sponsored by the non-profit organization, PKU Northwest. Participation was voluntary and confidentiality was guaranteed. The survey was answered anonymously with participants signing an informed consent form.

The researchers obtained approval from the Ethics Committee of the General Council of Official Associations of Psychologists (COP) in Spain. Additional approval was granted by the General Council of Psychology and the Vice-Rectorate for Scientific Policy and Research of Granada University, through a Contract Program between the Faculty of Education and Humanities of Melilla and the University of Granada in Spain.

2.2. Instrument

Data were obtained via the Questionnaire to Evaluate Knowledge of the Low Phe Diet in PKU, previously developed and validated by our team [28]. Completing the survey takes approximately 10 minutes. Participants were presented with 80 different foods and asked to determine whether each item could be eaten freely (without restriction or the need to measure the amount ingested) or not. We considered "free" those foods with a Phe content of less than 50 mg / 100g of product. The survey contained 35 "free" foods and 45 restricted items. A point is awarded for each correct response. The raw scores (number of points) yield a percentile score based on general knowledge as well as 3

percentile sub-scores measuring 1) knowledge of free foods (factor 1, reliability = .97), 2) easy to recognize restricted or prohibited foods (factor 2, reliability = .92) and 3) difficult to recognize limited foods (factor 3, reliability = .91). The Cronbach's alpha coefficient of the questionnaire is .945, indicating high internal reliability.

2.3. Statistical Analysis

Data were processed using SPSS Statistics 25 software. Descriptive statistics were used to synthesize and organize the data obtained. Quantitative data were presented as absolute frequency (n) and relative frequency (%) and shown in contingency tables. Quantitative variables are presented as position measures (mean and median) and dispersion (Standard Deviation and coefficients of variation). Non-parametrical tests (U of Mann Whitney or Kruskal Wallis) were used to evaluate significant differences between variables.

3. Results

A total of 207 participants answered the questionnaire. After eliminating those that included very incomplete forms, those that related to older patients or patients with benign forms that did not require treatment, data were analyzed from 172 surveys relating to 178 children with PKU. In total 37 states were represented in the study (Figure 1). The majority 11.2% were from California, 6.5% from Illinois and equal representation (6%) were from Washington, Ohio and Pennsylvania.

Caregivers were mostly women (86%) with a mean age of 40.6 years of age (SD=9.285). They were the mothers in 81.4% of cases; the father in 8.1% and 10.5% represented other caregivers. In our study population, 88.4% of the caregivers had completed university level studies, 8.1% had finished high school and only 2.9% had lower educational levels.

The 178 children with PKU they cared for were 49.4% boys and 50.6% girls with a mean age of 7.31 years (SD=4.628). 19.2% were 0-2 years of age, 23.9% were 3-5 years old, 27.9% were 6 - 10, 17.5% 11 -13 and 11.6% were 14- 17 years old. Severity of PKU was self-reported as classical in 87.2% of patients and moderate in 12.8%.

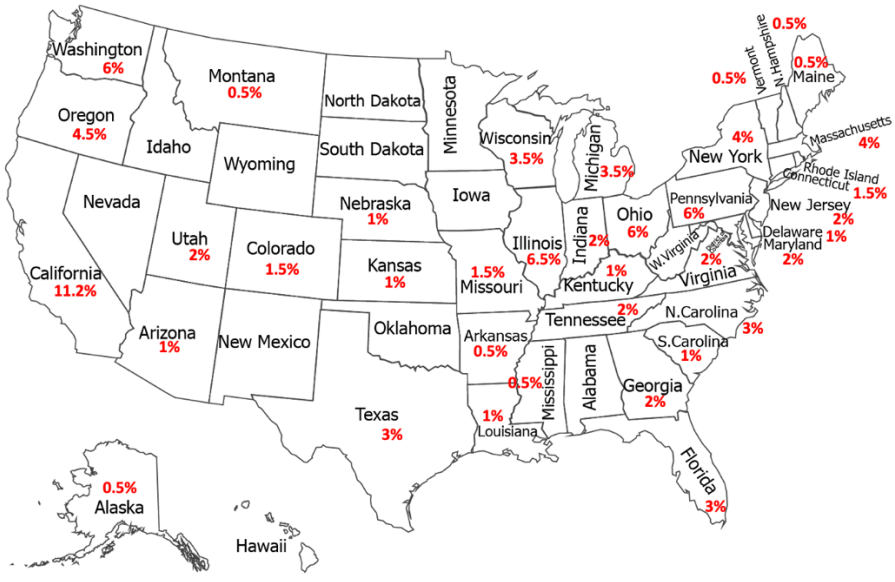

Figure 1. Geographical distribution of participants.

General knowledge of food selection among our caregivers was very high in 17.4% (70-99 score percentile), high in 42.4% (60-69 percentile), medium in 16.9% (percentile 40-59), low in 16.9% (percentile 30-39) and very low in 12.8% (0-29 percentile). Regarding specific food items, correct answers regarding easy to recognize restricted foods was between 98-99% (Table 1). However, among restricted foods not easy to recognize or allowed foods, correct answers varied from 40 to 60% (Table 2).

Table 1. Highest right answers.

Item food	% Wrong answers	Kind of food	Statistical standard deviation
Canned tuna	99	Not allowed	,108
Walnut	99	Not allowed	,108
Chicken	99	Not allowed	,108
Lobster	99	Not allowed	,108
Peanuts	99	Not allowed	,108
Cashew	99	Not allowed	,108
Egg	98	Not allowed	,131
Sausage	98	Not allowed	,131
Hazelnuts	98	Not allowed	,131
Whole-wheat bread	98	Not allowed	,131

Table 2. Highest wrong answers.

Item food	% Wrong answers	Kind of food	Statistical standard deviation
Saccharin	60.21	Allowed	,492
Mushrooms, white	59.83	Limited	,494
Artichoke	58.44	Limited	,496
Cocoa butter	57.02	Allowed	,497
Coconut	53.67	Limited	,501
Butter	55.23	Allowed	,499
Banana	46.65	Limited	,499
Pumpkin	45.81	Allowed	,498
Fresh orange juice	43.55	Allowed	,496
Avocado	40.46	Limited	,490

We found no statistically significant differences on food selection knowledge regarding the caregivers' sex, age or level of education. No differences were found in relation to the way the survey was answered (paper or online), or with the child characteristics (age, sex or PKU phenotype).

Caregivers were asked to self-report the number of blood tests to determine Phe controls they had sent in over the last 3 years and what percentage of those were above the concentration recommended by their physician. (23 respondents did not complete this section of the survey).

We found a significant correlation ($p<0.01$) between the food selection knowledge score and metabolic control (Pearson's r= .223), with lower scores associated with worse metabolic control. Respondents with lower scores were more likely to incorrectly label allowed foods (Pearson`s r= .252). We also found that younger caregivers and/or those with younger children reported worse metabolic control in their offspring (Pearson's r = .226).

4. Discussion

In summary, 3 out of 4 respondents demonstrated a medium, good or very good knowledge regarding food selection for their children with PKU. Participants were mainly mothers of young children with a high educational level and interested enough to belong to a PKU association and participate in an online survey. Consequently, they probably represent a highly motivated group and may be more likely to have high scores. A study including fathers and less well-educated respondents may not yield the same results. Moreover, the use of self-reports may have biased the results related to the number of blood tests submitted and the number of tests showing metabolic control above the recommended range. Despite this limitation, there was a significant correlation between the knowledge test score and this measure of metabolic control.

Although knowledge in general was relatively high, certain foods still posed a challenge for most respondents. These items were already identified during the validation process of the questionnaire and grouped within the "challenging" foods (factor 3) and they include mainly sauces or fruits and vegetables that have a high protein content. Regarding sauces such as mustard or mayonnaise no egg, these difficulties are probably due to the fact that their composition and therefore their Phe content can be highly variable and their classification as "free" or restricted is not always the same and can be confusing.

Also, some fruits such as avocado or vegetables such as artichoke contain higher concentrations of protein than most fruits or vegetables and therefore appear as limited foods in our questionnaire (Phe>50 mg for 100 gr of food). It is possible that in some PKU Centers they might be allowed and free, and in others they may be restricted.

Restricted foods were better identified than "free" foods. This result is not surprising, as educational efforts and the caregiver's attention is greater when avoiding those foods that might have a high Phe content that lead to high blood Phe levels. However, we believe that being able to identify foods that can be taken without restrictions is equally important in order to facilitate the planning and preparation of the diet, enhancing the introduction of a broader range of different foods, avoiding the sense of hunger and/or increasing the amount of calories, fiber or micronutrients from natural foods. In the long term, the broader and easier the diet, the more interesting it is and the more likely it is to enhance adherence to nutritional recommendations. This was particularly noted among the older cohorts of children. Adolescent with better metabolic control were those whose parents/caregivers had better general knowledge and more often correctly identified "free" foods.

The relatively large sample size and broad representation of respondents from throughout the United States suggests that this knowledge test is relevant for health professionals who work with families affected by classic and moderate PKU.

5. Conclusions

Having a measurement instrument to assess the general knowledge of the PKU diet has clinical utility. In particular, knowledge of allowed foods shows a significant correlation with the percentage of blood Phe tests exceeding recommended values. Future studies might include

socioeconomic variables as well as a broader range of participants who have less education or for whom English is not their native language.

Data also suggest that currently the focus of educational programs for parents or other caregivers has been on prohibited and limited foods rather than those allowed. Educational campaigns to increase awareness permitted foods in the PKU diet will increase the range and quality of foods offered to children and may improve long-term adherence to nutritional recommendations.

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