PAPER

# LIKING AND SENSORY DESCRIPTION OF PROTEIN SUBSTITUTES IN PHENYLKETONURIA SUBJECTS: A CASE-STUDY IN NORTHERN AND SOUTHERN ITALY

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## ABSTRACT

Nowadays, it is important to make effort to develop new formulations for subjects affected by rare diseases who need to follow a lifetime diet to maintain a good health.
The purpose of the study was to evaluate the acceptability and to obtain a sensory descriptive analysis of protein substitutes (glycomacropeptide GMP formulas *vs* L-amino acid formulas) involving subjects affected by phenylketonuria in Northern and Southern Italy. Results demonstrated in both groups of subjects a greater acceptability of GMP samples, characterized by sweet and mild taste, mild odor, and natural color, compared with amino acid formulations. These sensory attributes should be considered during product development as a key factor influencing subjects' satisfaction.

*Keywords*: acceptability, CATA, food development, food formulations, odor, taste

## 1. INTRODUCTION

Sensory perception and food preferences play a key role in determining food choice and, thus, directly influence the diet quality (COX *et al*., 2015). The study of the influence of sensory and hedonic individual differences in products acceptability is extremely important in modern food product development, especially for the ones meant to satisfy the needs of specific target populations (e.g, diabetics, elderly, obese etc). In this context, it is also important to make effort to develop new formulations for subjects affected by rare diseases who need to follow a lifetime diet to maintain a good state of health.
Between rare diseases, phenylketonuria (PKU) is an inborn metabolism error, which, if untreated, could lead to severe brain damage (GIOVANNINI *et al*., 2012). The main purpose of PKU patients' treatment is to control the blood phenylalanine (Phe) concentration to prevent severe health consequences (BLAU, 2016). The treatment for PKU is mainly a lifetime diet based on low-protein foods in combination with amino acid supplements. These supplements without Phe, are enriched with vitamins and minerals, and some products are also added with fat and carbohydrates to ensure normal growth and good health through adulthood (VAN SPRONSEN *et al*., 2017).
Compliance with this strict diet becomes a challenge over time, especially in adolescence, and this is primarily due to the unpleasant taste of the amino acid supplements and the variety of available formulations (MACDONALD *et al*., 2010, AGUIAR *et al*., 2015). In order to achieve better compliance with diet through the lifespan, protein supplements should satisfy the need for better taste and easier management (VAN SPRONSEN *et al*., 2008). The number of available protein substitutes for PKU subjects is increasing over time (FEILLET and AGOSTONI, 2010). In this context, beside the alternatives to traditional substitutes, the casein glycomacropeptide (GMP) is a 64-amino acid peptide from cheese whey which is rich in specific essential amino acids and is the only known natural protein source free from Phe (NEY *et al*., 2009; SOLVERSON *et al*., 2012).
The lack of empirical studies aimed at evaluating subjects' satisfaction for these substitutes reinforce the need to evaluate PKU subjects' perception regarding which sensory characteristics should have low-Phe formulations to be more appreciated. Only one recent study (PROSERPIO et a., 2018) has been conducted in an ambulatory context involving PKU subjects leading to define the sensory drivers of liking of protein substitutes in Northern Italy. However, intracultural differences between the Northern and Southern regions of Italy are well recognized, not only referred to industrialization and economic prosperity, but also to cultural values and social structures (RUGGIERO *et al*., 2000). These differences are also clearly reflected in different dietary patterns, which lead to a higher percentage of overweight and obese subjects in the Southern areas. In this context, some national data collected from the survey "Indagine Multiscopo dell'Istituto Nazionale di Statistica (ISTAT, 2015)" showed that, in the Southern regions, the consumption of food rich in carbohydrates prevails while healthy and low energy dense food consumption characterized the North West area.
Due of the above, the aim of the present study was to evaluate the acceptability of new GMP formulas compared with the more traditional amino-acid mixtures involving subjects affected by phenylketonuria in Northern and Southern Italy. A sensory descriptive analysis was also conducted in order to better understand which are the sensory characteristics that are related to the acceptance of these formulations that should be considered during the products development.

## 2. MATERIALS AND METHODS

### 2.1. Subjects

A total of sixty-six subjects (mean age: 25.6±5.9 years, 34 women and 32 men) gave informed consent and completed the study. Thirty-three subjects who were admitted to Department of Pediatrics, San Paolo Hospital (Milan, Italy), and thirty-three subjects referred to the Department of Pediatrics, Federico II University Hospital (Naples, Italy) were recruited. All participants were following a low-phenylalanine diet (metabolic control Phe (uMol/L): Milan= 642.1±309.2; Naples= 720.9±295.2). The exclusion criteria were: pregnancy, food allergies to whey proteins, severe neurological and functional disorders. The present study was performed according to the principles established by the Declaration of Helsinki after the protocol was approved by the Institutional Ethics Committee (protocol approval n°210).

### 2.2. Samples

4 L-amino acid formulas (AA) and 4 Glycomacropeptide (GMP) formulas, flavored with neutral, chocolate, strawberry and tomato aromas were prepared as reported in PROSERPIO *et al*., (2018).
The GMP flavored formulas (GMP_strawberry and GMP_tomato) were prepared by adding 2g of flavoring powder (aroMaxx erdbeere; aroMaxx tomate-basilikum, MetaX Istitut fur DiatetiK- MamoXi, Torino, Italy) to the neutral formulation (GMP_neutral) which consisted of 100 ml of Glytactin RTD™ (Cambrooke-Quaris, Roma, Italy). The GMP chocolate flavored sample (GMP_chocolate) consisted of 100 ml of Glytactin RTD™ Chocolate (Cambrooke-Quaris, Roma, Italy).
The AA neutral formula (AA_neutral) was prepared by mixing 16.5 g of powder high in L-amino acid (Xphe energy kid neutral, MetaX Istitut fur DiatetiK- MamoXi, Torino, Italy) and water to reach a final volume of 100 ml. The AA flavored formulas (AA_tomato; AA_chocolate) were prepared by adding 2g of flavoring powder (aroMaxx tomate-basilikum; aroMaxx schoko, MetaX Istitut fur DiatetiK- MamoXi, Torino, Italy). The L-amino acid strawberry flavored samples (AA_strawberry) were prepared using 16.5 g of Xphe energy kid erdbeere (MetaX Istitut fur DiatetiK- MamoXi, Torino, Italy) and water.
Each of these samples provides 5 g/100 ml protein equivalents. All the formulas were provided by MamoXi (Torino, Italy) and Cambrooke-Quaris (Roma, Italy).
30 ml of each sample were presented monadically, in a randomized and balanced order, to each participant in plastic cups labelled with three-digit codes. Water was available for rinsing the palate between the samples.

### 2.3. Experimental procedure

Sessions were conducted between 10:30-12:30 in quiet rooms under similar light conditions in both group of subjects (Milan and Naples). They were asked to refrain from consuming anything but water for 2 hours before the test (hungry state).
Subjects started by filling in the questionnaire on general appetite. Subsequently, they had to score their overall liking and, after a rest of 5 min, to made a sensory descriptive evaluation ('check-all-that-apply' questionnaire: CATA; VARELA and ARES, 2012) for each sample. All samples were prepared on the same day of the session and were presented at room temperature (20-22 °C).

### 2.4. General appetite

To ensure that both subjects from Milan and Naples were in a similar hunger state, they were asked to rate their appetite at the beginning of the session by filling out a questionnaire on general appetite (hunger, fullness, desire to eat, and thirst) all measured on 100mm Visual Analogue Scales (VAS, 'not at all': scored 0; 'very': scored 10).

### 2.5. Overall liking evaluation

Subjects were asked to taste the samples monadically and to express their liking scores on a 100mm VAS anchored by the extremes ''extremely disliked" (rated 0) and ''extremely liked" (rated 10). The experimenters provided instructions for the use of the scale prior to tasting (LAWLESS *et al*., 2010).

### 2.6. Sensory descriptive evaluation

The sensory descriptive evaluation was performed using the 'check-all-that-apply' (CATA) questionnaire. A separate group of 12 untrained subjects (age range: 20-40 years) attended a pilot test to define the suitable terms to describe the samples using a free listing questionnaire (VARELA and ARES, 2012). The eight low protein samples were provided to the subjects and they were asked to evaluate the sensory characteristics and to write all attributes for describing their color, appearance, odor, taste, flavor and texture. The individual development of the attributes was followed by an open discussion. Subsequently, the experimenters finalized the list of terms, selecting the most mentioned and the most common words in order to avoid synonymous (JAEGER *et al*., 2015).
Finally, the questionnaire consisted of a list of 27 sensory attributes: 10 for the appearance (light brown, dark brown, light yellow, dark yellow, light pink, dark pink, natural color, artificial color, brightness and opaque), 6 for the odor (natural odor, artificial odor, mild odor, strong odor, milk odor and vanilla odor), 8 for the taste/flavor (sweet, bitter, salty, sour, mild taste, strong taste, milk flavor and vanilla flavor) and 3 for the texture (thin, thick and floury).
Subjects who took part to the experimental sessions were asked to check from the list all the terms that they considered appropriate to describe each sample. The position of attributes was randomized.

### 2.7. Data analysis

A paired samples t-test was performed to compare the general appetite ratings (100mm VAS: hunger, fullness, satiety, desire to eat, and thirst) in the Northern and Southern groups before the samples evaluation.
A linear mixed model procedure was carried out on overall liking scores considering 'samples', 'gender' (women and men), 'city' (Northern and Southern) and their two-way interactions as fixed factors. Subjects were added as random effect while age as covariate. When a significant difference ($p<0.05$) was found, least significant difference (LSD) *post hoc* test was used. These statistical analyses were performed using IBM SPSS Statistics for Windows, Version 24.0 (IBM Corp., Armonk NY).
For the sensory descriptive evaluation, a data set was generated as 0/1 matrix, that is, "1" if the term was selected by the subjects and "0" if term was not selected. A frequency table was made from the total count of each term for each sample. Cochran's *Q* test was carried

out for each of the 27 terms to detect differences in participants' perception of the evaluated samples.
Correspondence analysis (CA) was used to obtain a bi-dimensional representation of the samples and to show relationship between samples and terms from the CATA questionnaire. Since the results provided by Northern and Southern subjects were similar to each other, the CA results are reported showing all the 66 subjects.
Penalty-lift analysis (PLAEHN, 2012; MEYNERS *et al*., 2013) was also performed in order to study which CATA terms were positively or negatively related with liking scores. These statistical analyses were performed using XLSTAT-Sensory® software for Windows, Version 2015.6.01 (Addinsoft™, France). A p-value of $<0.05$ was considered significant.

## RESULTS

### 3.1. General appetite

The baseline general appetite ratings (Table 1) confirmed that feelings of hunger, fullness, desire to eat, and thirst were not significantly different in the two groups of subjects (Northern and Southern).

**Table 1.** General appetite ratings (means±SEM), as measured on 100 mm VAS, provided by Northern and Southern subjects.

| General appetite | Northern | Southern | t | p |
|---|---|---|---|---|
| Hunger | 4.47±0.45 | 4.53±0.46 | 0.08 | 0.94 |
| Fullness | 3.43±0.43 | 3.53±0.49 | 1.69 | 0.09 |
| Desire to eat | 4.99±0.48 | 4.45±0.45 | 0.82 | 0.41 |
| Thirsty | 4.77±0.44 | 5.86±0.37 | 1.90 | 0.07 |

### 3.2. Overall liking evaluation

The main factor 'samples' was found to have a significant effect on liking scores ($F_{(7;441)}= 58,75$; $p<0.001$). Generally, GMP samples obtained significant higher liking scores (M=4.3±0.1) compared with the AA formulas (M=2.7±0.2).
A significant effect of the interaction 'samples' * 'city' on liking scores was also found ($F_{(7;441)}=2.95$, $p<0.01$). The mean liking scores by samples in subjects from Milan and Naples are provided in Table 2.
Looking at the results gave by Northern subjects, the highest liking scores were obtained by the GMP_chocolate and GMP_strawberry, which were comparable to each other. The AA_strawberry samples obtained the highest score between the AA formulas, even if all AA samples were not acceptable (score <5). Similarly, Southern subjects preferred the GMP and AA samples flavored with strawberry aroma. Contrariwise, the tomato flavored samples, both GMP and AA formulas, obtained the lowest hedonic ratings in both subjects from Milan and Naples.

**Table 2.** Liking scores (means±SEM) for each samples provided by Northern and Southern subjects. Different letters (in column) indicate significant differences according to *post hoc* test within each group of subjects.

| Samples | | Liking scores | | |
|---|---|---|---|---|
| | | Northern | Southern | p |
| GMP | tomato | $1.10^{a}$±0.38 | $1.23^{a}$±0.39 | 0.78 $^{n.s}$ |
| | neutral | $4.48^{c}$±0.37 | $4.45^{cd}$±0.39 | 0.89 $^{n.s}$ |
| | chocolate | $5.64^{d}$±0.37 | $4.84^{d}$±0.38 | 0.13 $^{n.s}$ |
| | strawberry | $6.45^{d}$±0.38 | $6.13^{e}$±0.40 | 0.57 $^{n.s}$ |
| AA | tomato | $1.17^{a}$±0.38 | $0.94^{a}$±0.38 | 0.72 $^{n.s}$ |
| | neutral | $1.23^{a}$±0.37 | $3.10^{b}$±0.39 | 0.00 *** |
| | chocolate | $3.00^{b}$±0.38 | $3.33^{bc}$±0.39 | 0.55 $^{n.s}$ |
| | strawberry | $4.05^{c}$±0.37 | $5.17^{de}$±0.38 | 0.03* |

*p<0.05; *** p<0.001.

Comparing the results obtained in the two groups of subjects no significant differences have been highlighted between the GMP samples, while significant differences have been found in AA samples. In particular, significant higher scores were provided to AA_neutral and AA_strawberry by Southern subjects.
The main factor 'gender' and the interactions 'sample' * 'gender'; 'city' * 'gender' were not significant ($F_{(1,64)}$= 0.015, p=0.90; $F_{(7,441)}$= 0.70, p=0.80; $F_{(1,64)}$=0.004, p=0.95, respectively).

### 3.3. Sensory descriptive evaluation

The contingency table below (Table 3) shows the frequency of terms checked by Northern and Southern subjects to describe the eight samples.
Significant differences (p<0.001) were found in the frequency for all the 27 terms within the five sensory attributes categories evaluated, suggesting that the PKU subjects involved perceived differences between samples in terms of their sensory characteristics.
The Correspondence Analysis, used to obtain a bi-dimensional representation of the samples and the relationship between samples and terms from the CATA questionnaire, resulted in two dimensions accounting for 60.62% of variance in the data.
As shown from Fig. 1, samples were clearly discriminated by subjects according to their aroma. Indeed, samples flavored with strawberry aroma (AA_strawberry and GMP_strawberry) are positioned in the upper left side of the map while the samples added with tomato aroma (AA_tomato and GMP_tomato) are situated in the upper right side of the map. In the lower part of the map, GMP samples without aroma (GMP_neutral) and the chocolate one (GMP_chocolate) are well distinguished from the L-amino acid formulas with the same aromas (AA_neutral and AA_chocolate).

**Table 3.** Contingency table for the sensory descriptive analysis evaluation.

| Sensory attributes | AA samples | | | | GMP samples | | | |
|---|---|---|---|---|---|---|---|---|
| | Chocolate | Strawberry | Neutral | Tomato | Chocolate | Strawberry | Neutral | Tomato |
| *Appearance* | | | | | | | | |
| artificial color | 9 | 28 | 12 | 20 | 10 | 24 | 14 | 26 |
| natural color | 17 | 7 | 24 | 6 | 19 | 7 | 25 | 7 |
| light yellow | 0 | 0 | 5 | 1 | 12 | 0 | 19 | 1 |
| dark yellow | 0 | 0 | 0 | 3 | 12 | 0 | 1 | 7 |
| brightness | 2 | 18 | 15 | 4 | 11 | 5 | 12 | 5 |
| light brown | 5 | 0 | 0 | 45 | 25 | 0 | 3 | 35 |
| dark brown | 60 | 0 | 0 | 7 | 4 | 0 | 0 | 4 |
| opaque | 19 | 3 | 6 | 18 | 13 | 13 | 16 | 23 |
| light pink | 0 | 58 | 0 | 0 | 0 | 15 | 0 | 4 |
| dark pink | 0 | 2 | 0 | 6 | 0 | 47 | 0 | 9 |
| *Odor* | | | | | | | | |
| artificial odor | 21 | 25 | 32 | 34 | 12 | 23 | 10 | 35 |
| mild odor | 15 | 38 | 19 | 2 | 26 | 24 | 27 | 6 |
| milk odor | 4 | 4 | 8 | 0 | 14 | 6 | 32 | 2 |
| vanilla odor | 1 | 7 | 13 | 1 | 13 | 4 | 11 | 2 |
| strong odor | 21 | 5 | 11 | 43 | 10 | 19 | 7 | 40 |
| natural odor | 20 | 14 | 8 | 3 | 20 | 9 | 18 | 3 |
| *Taste* | | | | | | | | |
| sweet | 14 | 39 | 6 | 1 | 41 | 51 | 33 | 5 |
| sour | 10 | 12 | 17 | 26 | 2 | 3 | 6 | 23 |
| salty | 12 | 6 | 15 | 38 | 3 | 0 | 5 | 31 |
| bitter | 27 | 9 | 28 | 24 | 8 | 1 | 6 | 18 |
| mild taste | 11 | 27 | 18 | 2 | 31 | 27 | 32 | 5 |
| strong taste | 35 | 20 | 28 | 51 | 7 | 23 | 9 | 45 |
| *Flavor* | | | | | | | | |
| milk flavor | 6 | 9 | 2 | 0 | 24 | 10 | 35 | 1 |
| vanilla flavor | 1 | 4 | 8 | 0 | 16 | 3 | 8 | 2 |
| *Texture* | | | | | | | | |
| thin | 18 | 44 | 39 | 29 | 24 | 25 | 32 | 25 |
| thick | 32 | 0 | 17 | 16 | 18 | 21 | 20 | 25 |
| floury | 21 | 2 | 14 | 12 | 11 | 8 | 6 | 8 |

## 3.4. Relating sensory profiling with liking

A penalty-lift analysis was carried out to understand which sensory attributes were mainly associated to the overall liking of the samples. The analysis showed to which extent liking increased or decreased when the subjects related a certain CATA terms to the samples.

As inferred from Fig. 2 liking scores were significantly positively associated with the sensory attributes: 'sweet' ($p<0.0001$), 'mild taste' ($p<0.0001$), 'mild odor' ($p<0.0001$) and

'natural color' ($p<0.0001$). Contrariwise, the terms that significantly decreased the samples' acceptability were: 'salty' ($p<0.0001$); 'strong taste' ($p<0.0001$); 'bitter' ($p<0.0001$); 'strong odor'($p<0.0001$); 'light brown'($p<0.0001$); 'artificial odor'($p<0.0001$), 'opaque'($p<0.001$) and 'thick'($p<0.05$).

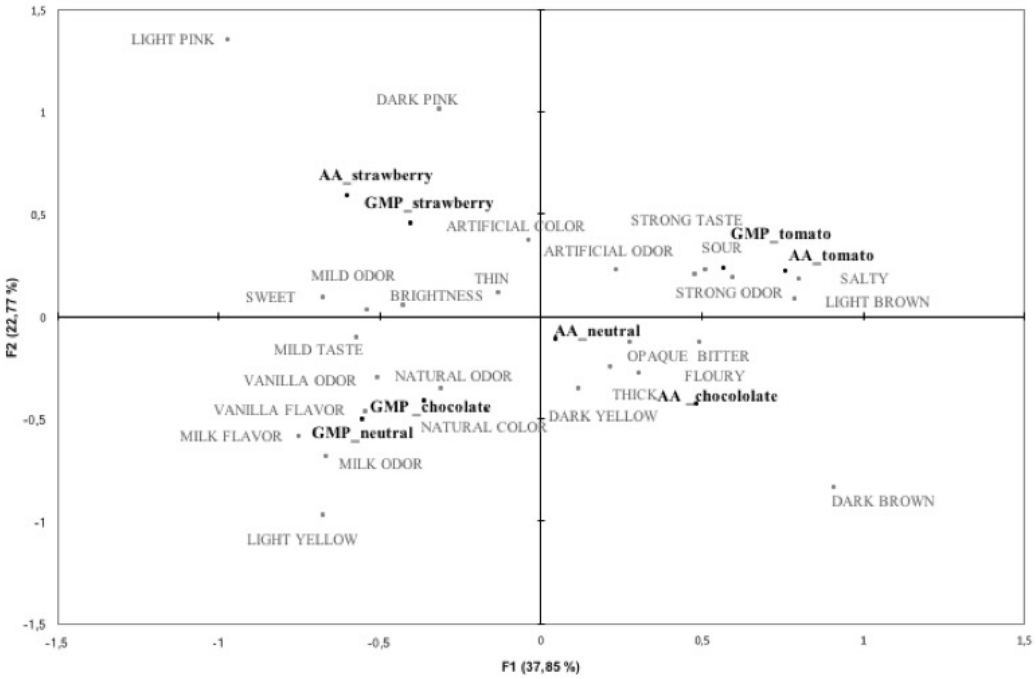


**Figure 1.** Attributes and products plot obtained from CATA total frequency counts.

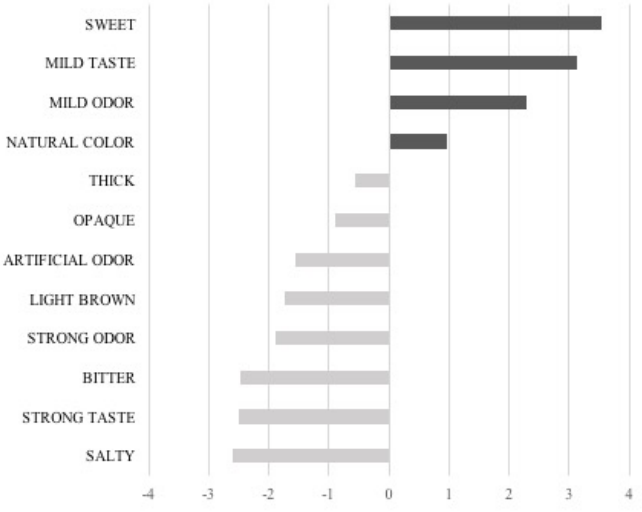


**Figure 2.** Penalty-lift analysis of sensory attributes across all samples.

## 4. DISCUSSION

The purpose of the present study was to deepen the evaluation of the acceptability of protein substitutes (GMP formulas *vs* L-amino acid formulas) involving subjects affected by phenylketonuria in Northern and Southern Italy. A sensory descriptive evaluation was performed to define which sensory characteristics are mainly related to the acceptance or to the refusal of these products that should be taken into account in developing new formulations for specific target populations.

Overall, the present results demonstrated in both subjects from Milan and Naples a greater acceptability of GMP samples compared with the more traditional amino acid formulations, besides the reported differences in dietary pattern among the regions of Italy (RUGGIERO *et al*., 2000; ISTAT, 2015). In particular, GMP samples flavored with chocolate and strawberry, were the most appreciated by the Northern group, while the sample flavored with strawberry was the most appreciated by the Southern group. These results confirmed our previous findings with a group of Northern subjects (PROSERPIO *et al*., 2018) in which a greater acceptability was depicted for GMP beverages flavored with chocolate and strawberry compared with the L-amino acid formulations flavored with the same aromas. According to the present results Lim and collaborators (2007) showed that a GMP chocolate beverage was significantly more liked compared with the same flavored amino acid beverage.

Both groups of subjects from Milan and Naples gave to the tomato flavored samples the lowest liking scores. Even if the GMP samples received generally average higher liking scores (M=4.3±0.1) compared with the AA formulas (M=2.7±0.2), it is important to consider that the type of added aroma was a key driver of the acceptance. Indeed, the GMP sample, as well as AA formulation, flavored with the tomato aroma were evaluated as not acceptable by both group of subjects. These samples were not appreciated since they were perceived as salty, sour and characterized by strong odor and taste. The tomato flavored formulations maybe were perceived as unpleasant since tomato aroma, signaling savory product, is not usually used as flavoring in low-phenylalanine products, especially as beverages. Thus, subjects maybe have perceived tomato flavored samples as really far from their usual food habits. Indeed, it is well known that food habits and also consumers' expectations could influence the sensory perception, the liking, and consequently the actual food consumption (KÖSTER, 2009). Confirming this hypothesis, two different studies demonstrated that cracker samples with GMP, a product expected to be characterized by salty taste, obtained higher liking scores compared with the low-protein crackers (LIM *et al*., 2007; VAN CALCAR *et al*., 2012).

Considering the sensory attributes mainly associated to the overall liking of the samples, the present study demonstrated that the acceptability increased when the samples were characterized by the attributes: 'sweet', 'mild taste', 'mild odor' and 'natural color'. The information achieved by the present results could be useful to understand which sensory characteristic should have a low-phenylalanine product to be more accepted by the subjects. Consequently, more appreciated products could facilitate dietary compliance that is a challenge over time, especially in adolescence (MACDONALD *et al*., 2010, AGUIAR *et al*., 2015). Moreover, it has also been demonstrated that, besides the higher acceptance of GMP formulations compared the more traditional ones, GMP could also be considered as a more physiological source of dietary protein and promote higher satiety compared with synthetic amino acids (VAN CALCAR *et al*., 2012).

As future perspective, it should be useful to evaluate other GMP base products using validated sensory approaches, as it has been performed in the present study, besides considering mainly the nutritional composition of these products.
In conclusion, the present results suggest that, in order to improve the protein substitutes' sensory quality, these formulations should be characterized by a sweet and mild taste, a mild odor, and a natural color. Indeed, it is important to consider the PKU patients' satisfaction as a key factor during the product development in order to improve the diet quality through the lifespan.

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