

**БИОТЕХНОЛОГИЯ ПРОДУКТОВ ПИТАНИЯ И БИОЛОГИЧЕСКИХ АКТИВНЫХ ВЕЩЕСТВ /
BIOTECHNOLOGY OF FOOD AND BIOLOGICAL ACTIVE SUBSTANCES**

DOI: <https://doi.org/10.23649/JAE.2024.42.2>

DEVELOPMENT OF MUFFIN FORMULATIONS WITH LOW PHENYLALANINE CONTENT

Research article

Bichenkova V.V.^{1,*}, Bazhenova I.A.², Chernova E.V.³, Kotova N.P.⁴

¹ORCID : 0000-0002-4807-1612;

³ORCID : 0000-0001-8322-9027;

^{1,2} St. Petersburg Polytechnic University, Saint-Petersburg, Russian Federation

³ Saint-Petersburg State University of Economics, Saint-Petersburg, Russian Federation

⁴ College of Culinary Skills, Saint-Petersburg, Russian Federation

* Corresponding author (lera_bychenkova[at]mail.ru)

Abstract

Human quality of life is not least determined by nutrition. Therefore, scientists and practical workers face a problem to create specialized products for people suffering from various diseases, including genetic ones. This paper presents the experience gained by the scientists of the Higher School of Biotechnology and Food Production, Peter the Great Polytechnic University, in developing a technology for producing low-protein muffins for patients with phenylketonuria. Wheat flour formulation served as the control sample. In the formulations developed, wheat flour was replaced by rice flour. Banana puree and orange juice (20% of the total weight each); sodium pyrophosphate (E450) and sodium hydrogen carbonate (E500) in quantities not exceeding 1% of the semi-finished product weight were used to improve the structural and mechanical properties of the product, upgrade the quality of the finished product and its biological value. Several product formulations were developed with different ratios of rice flour and corn starch. The optimal sample in terms of phenylalanine amount and organoleptic characteristics was found to be the one with the ratio of rice flour to corn starch 1:1. The phenylalanine content in this sample was 7.5 times lower compared to the control sample.

Keywords: phenylketonuria, special nutrition, flour confectionery goods.

РАЗРАБОТКА РЕЦЕПТУР МАФФИНОВ С НИЗКИМ СОДЕРЖАНИЕМ ФЕНИЛАЛАНИНА

Научная статья

Быченкова В.В.^{1,*}, Баженова И.А.², Чернова Е.В.³, Котова Н.П.⁴

¹ORCID : 0000-0002-4807-1612;

³ORCID : 0000-0001-8322-9027;

^{1,2} Санкт-Петербургский Политехнический университет, Санкт-Петербург, Российская Федерация

³ Санкт-петербургский государственный экономический университет, Санкт-Петербург, Российская Федерация

⁴ Колледж кулинарного мастерства, Санкт-Петербург, Российская Федерация

* Корреспондирующий автор (lera_bychenkova[at]mail.ru)

Аннотация

Качество жизни человека не в последнюю очередь определяется питанием. Поэтому перед учеными и практиками стоит задача создания специализированной продукции для людей, страдающих различными заболеваниями, в том числе генетическими. В статье представлен опыт ученых Высшей школы биотехнологии и пищевых производств Политехнического университета Петра Великого при разработке технологии производства низкобелковых сдобы для больных фенилкетонурией. Контрольным образцом служила рецептура кекса из пшеничной муки. В разработанных рецептурах кексов пшеничная мука заменена рисовой. Банановое пюре и апельсиновый сок (по 20% от общей массы каждого); пирофосфат натрия (E450) и гидрокарбонат натрия (E500) в количествах, не превышающих 1% от массы полуфабриката, использовали для улучшения структурных и механических свойств продукта, повышения качества готового продукта и его биологической ценности. Было разработано несколько рецептур изделий с различным соотношением рисовой муки и кукурузного крахмала. Оптимальным по количеству фенилаланина и органолептическим показателям оказался образец с соотношением рисовой муки и кукурузного крахмала 1:1. Содержание фенилаланина в этом образце было в 7,5 раза ниже по сравнению с контрольным образцом.

Ключевые слова: фенилкетонурия, специальное питание, мучные кондитерские изделия.

Introduction

Scientists in the Russian Federation pay great attention to the development of special nutrition for people suffering from various genetic and alimentary-dependent diseases. The authors of the article developed products and dishes for specialized nutrition [1], worked on the creation of products and dishes of increased biological value [2], [3], studied Russian cuisine from the point of view of functional nutrition [4], [5].

At present, a part of the population in the Russian Federation has phenylketonuria. "oPPhenylketonuria is a hereditary disease caused by a deficiency in the activity of phenylalanine hydroxylase, which converts phenylalanine to tyrosine" [6], resulting in the formation of toxic substances [7, P. 33]. Excess phenylalanine and products of its metabolism lead to damage to the central nervous system up to mental retardation [8]. There is no therapy to treat this disease. The only precondition for normal functioning of the body is a lifelong low-protein diet [9], [10]. Phenylketonuria severely limits food choices and

consists in the inability to consume animal protein [11]. Researchers state that "with timely detection of the disease (neonatal screening) and subsequent treatment, symptoms of the disease are milder or absent. Nutritional therapy should be started no later than the first weeks of the child's life" [12].

Methodical recommendations present a list of products recommended and prohibited in the diet of patients, and the permissible quantity of phenylalanine in the diet of children and adults [9, P. 25-30, P. 59-62]. In Russia, as in other countries, the division of products is based on the principle of traffic lights [13, P. 31-32], [14], [15, P. 23-24].

In order to expand the range of protein products in the diet of patients with phenylketonuria, physicians suggest to use drug therapy [16] and diet [17], [18]. The need for non-drug therapy through specialized therapeutic products is increasing [11], [19]. Scientists of the Republic of Belarus have developed pasta, porridge mixes [20], [21]. However, the number of products for patients with phenylketonuria does not differ in a wide range, including there are no flour confectionery products.

The study objective was to select raw materials and develop a formulation for a low-protein flour confectionery product, which can be recommended for the nutrition of patients with phenylketonuria.

Study methods and principles

The objects of the study were:

- muffins produced according to the traditional formulation based on wheat flour (control sample);
- muffins for special nutrition, produced on the basis of mixtures of rice flour and corn starch in different ratios.

The following raw materials were used in the production of muffins cooked according to traditional and developed formulations: straight white wheat flour, rice flour, corn starch, sugar sand, melange, vegetable oil, orange juice, banana puree, sodium hydrogen carbonate raising agent. Muffin cooking technology: pre-prepared raw materials were combined with fruit puree, orange juice and vegetable oil and kneaded to a smooth emulsion with a Braun MQ 535 blender. The products were molded in silicone baking molds. They were baked in an Electrolux EZB52410AW convection oven at 180°C for 25 minutes. The finished products were cooled for 15-20 minutes at 18-20°C.

Study methods: moisture content was determined by drying method according to GOST 5900-2014 [22], alkalinity was assessed by titration method according to GOST 5898-87 [23]. The density index and the water absorption index are according to GOST 15810-2014 [24]. The vitamins and minerals content, energy value were determined by calculation methods. Phenylalanine content was determined according to the "PKU Diet" program [25].

Results and discussion

When selecting raw materials for patients with phenylketonuria, special attention should be paid to the phenylalanine content in them. Methodological manuals on nutritional management for patients in this group indicate that "the recommended ratios of the main nutrients in foods are set differently depending on the age groups of patients" [26]. The permissible daily phenylalanine quantity for children over 6 years of age is 15-10 mg/kg body weight [27], the range of phenylalanine consumption is 290-1,200 mg/day for men over 19 years of age and 220-700 mg/day for women over 19 years of age [28]. The paper analyzes the objects of research. Corn and rice flour, corn and rice starch are permitted products for patients with phenylketonuria [9, P. 61]. Ayrumyan V.Yu. et al. refer rice flour to dietary products [29], and the Ministry of Health of the Russian Federation recommends the use of corn starch to create low-protein products [26, P. 21]. The phenylalanine content in 100 grams of these products ranges from 19 mg to 380 mg in corn flour [30]. It is impossible to completely eliminate phenylalanine from raw materials, because protein is an important component that determines the structural and mechanical properties of flour products [30]. Table 1 shows the ratio of the main components of the formulation in the studied samples.

Table 1 - Ratio of components in samples

DOI: <https://doi.org/10.23649/JAE.2024.42.2.1>

Components of the mixture	Quantity, %		
	sample 1	sample 2	sample 3
Rice flour	100	70	50
Corn starch	-	30	50

It was found (Based on a comparison of water density and absorption indicators for products with different ratios of rice flour and corn starch) that corn starch in the amount of up to 50% relative to rice flour could be used without deterioration of quality of the finished products. The recipe of the cupcake of sample 3 is shown in the table 2.

Table 2 - Cupcake recipe

DOI: <https://doi.org/10.23649/JAE.2024.42.2.2>

Products	Mass fraction of dry substances, %	Consumption of products per 100 pieces of finished products, g	
		in kind	in dry substances
Rice flour	88,00	1580	1390,4
Corn starch	87,00	1580	1374,6
Sugar	99,85	1765	1762,4

Orange juice	13,00	1860	241,8
Banana puree	26,00	1860	483,6
Vegetable oil	99,99	555	554,9
Baking powder	95,00	90	85,5
Total	-	9290	5893,2
Exit	74,3	7500	5569,1
Humidity 25,7 ± 2,0%		-	

Note: sample 3

Nutritional and energy values of products are presented in the table 3.

Table 3 - Nutritional and energy value of muffins

DOI: <https://doi.org/10.23649/JAE.2024.42.2.3>

Name	Proteins, g	Fats, g	Carbohydrates, g	Energy value, kcal
Control sample	4.0	8.0	40.5	250
Sample 1	2.3	7.1	40.0	231
Sample 2	1.5	7.0	39.5	227
Sample 3	1.1	6.9	38.0	218

Experimental formulations used gluten-free flour. Banana puree and orange juice (20% of the total weight each); sodium pyrophosphate (E450) and sodium hydrogen carbonate (E500) in quantities not exceeding 1% of the semi-finished product weight were used to improve the properties of the dough, to upgrade the quality of the finished product and its biological value. The choice of banana puree and orange juice for use in the formulation is justified by the fact that the diet of patients with phenylketonuria is characterized by a deficiency of vitamins and mineral elements, which must be compensated [28]. The addition of these ingredients reduced the amount of sugar in the formulation and decreased the caloric value from 250 to 218 kilocalories per 100 grams of finished product. In addition, banana puree is a structure-forming agent due to pectin substances. The third sample was not inferior to the control in taste, color, texture, and odor, while the parameter "appearance" was lower than the control by only 0.5 points. The phenylalanine content in the control sample was 434 mg/100 g of the finished product, in the developed products it was as follows: No. 1 – 99, No. 2 – 75, No. 3 – 56.5 mg/100 g of the finished product. In the third sample, the phenylalanine content was 7.5 times lower as compared to the control sample.

According to the study results, it was concluded that the optimal formulation for the production of low-protein muffins was formulation number three. The products obtained according to this formulation have good organoleptic characteristics, low phenylalanine and carbohydrate content. Physico-chemical parameters of the muffin with low protein and phenylalanine content: moisture content – 21.98±0.84%, alkalinity – 0.310±0.03 deg. The developed product does not differ from the control version in appearance and organoleptic characteristics.

Conclusion

Attention should be paid to the choice of raw materials in the production of specialized products for patients with phenylketonuria. The use of wheat flour for people with this disease is limited. In the developed products, wheat flour was replaced by rice flour and corn starch. Analysis of quality characteristics of the developed products showed that the experimental samples were not inferior to the control by organoleptic and physico-chemical parameters, met the standards for the phenylalanine content allowed in the diet of patients of this category. The quantity of phenylalanine in the developed product is 56.5 mg/100 g of the finished product, which is 7.5 times less than that in the muffin made using wheat flour.

The developed product has low content of protein (1.1 g/100 g) and phenylalanine (56.5 mg/100 g), expanding the range of products for patients with phenylketonuria, which contributes to improving their quality of life.

Конфликт интересов

Не указан.

Рецензия

Все статьи проходят рецензирование. Но рецензент или автор статьи предпочли не публиковать рецензию к этой статье в открытом доступе. Рецензия может быть предоставлена компетентным органам по запросу.

Conflict of Interest

None declared.

Review

All articles are peer-reviewed. But the reviewer or the author of the article chose not to publish a review of this article in the public domain. The review can be provided to the competent authorities upon request.

Список литературы / References

1. Bazhenova I.A. Germination of Proso Millet (*Panicum miliaceum* L.) Grains Trigger Biochemical Changes That Augment Bioavailability of Flower and Its Utility for Gluten-free Dietary Foods / I.A. Bazhenova, T.S. Bazhenova, E.V. Chernova // *Agronomy Research*. — 2019. — Vol. 17. — № 6. — P. 2187-2194.
2. Чернова Е.В. Новый метод оценки биологической ценности белков кулинарно обработанных круп / Е.В. Чернова // *Известия высших учебных заведений. Пищевая технология*. — 2001. — № 1 (260). — С. 11-13.
3. Chernova E. Development of the Composition of Cereal Dishes of Higher Biological Value / E. Chernova, I. Bazhenova, T. Bazhenova // *BIO Web of Conferences*. — 2021. — Vol. 29. — Art. Number 01022.
4. Чернова Е.В. Русская национальная кухня и функциональное питание / Е.В. Чернова, В.Н. Красильников // *Пищевая промышленность*. — 2001. — № 8. — С. 64-65.
5. Баженова Т.С. Применение пшеничной муки в производстве безглютенового бисквита / Т.С. Баженова, И.А. Баженова // *XXI век: итоги прошлого и проблемы настоящего плюс*. — 2018. — Т. 7. — № 2 (42). — С. 39-42.
6. Wegberg A.M.J. The Complete European Guidelines on Phenylketonuria: Diagnosis and Treatment / A.M.J. Wegberg, A. MacDonald, K. Ahring [et al.] // *Orphanet J Rare Dis*. — 2017. — Vol. 12. — № 1. — P. 162.
7. Хальчицкий С.Е. Исследование нарушений метаболизма фенилаланина и его дериватов при наследственных и экзогенных гиперфенилаланиниемиях: дис. ... канд. биол. наук / С.Е. Хальчицкий. — СПб., 2017. — 162 с.
8. Бушуева Т.В. Современный взгляд на проблему фенилкетонурии у детей: диагностика, клиника, лечение / Т.В. Бушуева // *Вопросы современной педиатрии*. — 2010. — № 9 (11). — С. 57-162.
9. Клинические рекомендации по диагностике и лечению фенилкетонурии и нарушений обмена тетрагидробиоптерина. — М.: Академиздат, 2014. — 70 с.
10. Wood G. Special Low Protein Foods Prescribed in England for PKU Patients: An Analysis of Prescribing Patterns and Cost / G. Wood, Sh. Evans, A. Daly [et al.] // *Nutriens*. — 2021. — Vol. 13. — № 11. — Art. Number 3977.
11. Боровик Т.Э. Диетотерапия при классической фенилкетонурии: критерии выбора специализированных продуктов без фенилаланина / Т.Э. Боровик, К.С. Ладодо, Т.В. Бушуева [и др.] // *Вопросы современной педиатрии*. — 2013. — № 12 (5). — С. 40-48.
12. Клиточенко Г.В. Современные возможности диагностики и терапии фенилкетонурии / Г.В. Клиточенко, Н.В. Малюжинская, К.В. Степаненко // *Лекарственный вестник*. — 2021. — Т. 15. — № 1 (81). — С. 24-29.
13. Классическая фенилкетонурия и другие виды гиперфенилаланинемии: клинические рекомендации. — 2020. — 112 с.
14. Корнева О.А. Исследование кривой вязкости пресного безглютенового теста в зависимости от концентрации разных видов муки / О.А. Корнева, Е.Г. Дунец, О.В. Руденко [и др.] // *Пищевая промышленность*. — 2019. — № 6. — С. 62-65.
15. Бушуева Т.В. Специализированные продукты лечебного питания для детей с фенилкетонурией / Т.В. Бушуева [и др.] — М.: НМИЦ здоровья детей, 2018. — 128 с.
16. Николаева Е.А. Диагностика и лечение биоптериндефицитной гиперфенилаланинемии / Е.А. Николаева, М.И. Яблонская, М.Н. Харабадзе [и др.] // *Российский вестник перинатологии и педиатрии*. — 2015. — № 2. — С. 66-71.
17. Spronsen F. Key European guidelines for the diagnosis and management of patients with phenylketonuria / F. Spronsen, A. Wegberg, K.K. Ahring [et al.] // *The Lancet Diabetes & Endocrinology*. — 2017. — Vol. 5(9).
18. Vockley J. Phenylalanine hydroxylase deficiency: diagnosis and management guideline / J. Vockley, H.C. Andersson, K.M. Antshel [et al.] // *Genet. Med*. — 2014. — Vol. 16. — № 2. — P. 188-200.
19. Семенова Н.А. Исследование эффективности продуктов сухих специализированных для диетического лечебного питания детей, взрослых и беременных женщин, больных фенилкетонурией / Н.А. Семенова, Г.В. Байдакова, Н.В. Никитина [и др.] // *РМЖ. Мать и дитя*. — 2019. — Т. 2. — № 4. — С. 355-360.
20. Мадзиевская Т. Новые смеси для производства специализированных макаронных изделий / Т. Мадзиевская, Т. Шункевич, А. Белая // *Наука и инновации*. — 2014. — Т. 5. — № 135. — С. 42-43.
21. Ловкис З.В. Разработка низкобелковых макаронных изделий для питания людей с нарушением обмена фенилаланина / З.В. Ловкис, А.В. Садовская, Ю.С. Усеня [и др.] // *Пищевая промышленность: наука и технологии*. — 2020. — Т. 13. — № 3 (49). — С. 6-11.
22. ГОСТ 5900-2014. Изделия кондитерские. Методы определения влаги и сухих веществ. — М.: Стандартинформ, 2019. — 10 с.
23. ГОСТ 5898-87 Изделия кондитерские. Методы определения кислотности и щелочности. — М.: Стандартинформ, 2012. — 26 с.
24. ГОСТ 5897-90 Изделия кондитерские. Методы определения органолептических показателей качества, размеров, массы нетто и составных частей. — М.: Стандартинформ, 2012. — 16 с.
25. Автоматизированный расчет диеты. Фенилкетонурия // Диета ФКУ. — URL: http://pku.asteis.net/help_2.html#diet (дата обращения: 12.09.2023).
26. Специализированные продукты лечебного питания для детей с фенилкетонурией: Методическое письмо / Сост. А.А. Баранов, Т.Э. Боровик, К.С. Ладодо [и др.] — М., 2012. — 84 с.
27. Рылова Н.В. Нарушение обмена фенилаланина / Н.В. Рылова. — URL: http://www.rusnauka.com/31_NNM_2013/Medicine/5_143828.doc.htm. (дата обращения: 12.09.2023).
28. MacLeod E. Nutritional Management of Phenylketonuria / E. MacLeod, D.M. Ney // *Annals of Nutrition and Metabolism*. — 2010. — Vol. 68(2). — P. 58-69.
29. Содержание фенилаланина в основных продуктах питания // Вместе со всеми (Форум «Все о фенилкетонурии»). — URL: <https://clck.ru/38VTh6> (дата обращения: 12.09.2023).

30. Soltanizadeh N. Strategies Used in Production of Phenylalanine-Free Foods for PKU Management / N. Soltanizadeh, L. Mirmoghtadaie // *Comprehensive Reviews in Food Science and Food Safety*. — 2014. — Vol. 13. — № 3. — P. 287-299.
31. Иванова З.А. Разработка технологии хлебобулочных изделий с использованием рисовой муки / З.А. Иванова, Ф.Х. Тхазеплова, И.Б. Шогенова // *Проблемы развития АПК региона*. — 2018. — № 3 (35). — С. 168-170.
32. Айрумян В.Ю. Химический состав продуктов переработки зерна риса и кукурузы для повышенной пищевой и биологической ценности хлебобулочных изделий / В.Ю. Айрумян, Н.В. Сокол, Е.А. Ольховатов // *Ползуновский вестник*. — 2020. — № 3. — С. 3-10.

Список литературы на английском языке / References in English

1. Bazhenova I.A. Germination of Proso Millet (*Panicum miliaceum* L.) Grains Trigger Biochemical Changes That Augment Bioavailability of Flower and Its Utility for Gluten-free Dietary Foods / I.A. Bazhenova, T.S. Bazhenova, E.V. Chernova // *Agronomy Research*. — 2019. — Vol. 17. — № 6. — P. 2187-2194.
2. Chernova E.V. Novyj metod ocenki biologicheskoy cennosti belkov kulinarno obrabotannykh krup [A New Method of Estimation of Biological Value of Proteins of Cooking Processed Cereals] / E.V. Chernova // *Izvestija vysshix uchebnykh zavedenij. Pishhevaja tehnologija* [Proceedings of Higher Educational Institutions. Food Technology]. — 2001. — № 1 (260). — P. 11-13. [in Russian]
3. Chernova E. Development of the Composition of Cereal Dishes of Higher Biological Value / E. Chernova, I. Bazhenova, T. Bazhenova // *BIO Web of Conferences*. — 2021. — Vol. 29. — Art. Number 01022.
4. Chernova E.V. Russkaja nacional'naja kuhnja i funkcional'noe pitanie [Russian National Cuisine and Functional Nutrition] / E.V. Chernova, V.N. Krasil'nikov // *Pishhevaja promyshlennost'* [Food Industry]. — 2001. — № 8. — P. 64-65. [in Russian]
5. Bazhenova T.S. Primenenie pshennoj muki v proizvodstve bezglutenovogo biskvita [Application of Millet Flour in the Production of Gluten-Free Biscuit] / T.S. Bazhenova, I.A. Bazhenova // *XXI vek: itogi proshlogo i problemy nastojashhego pljus* [XXI Century: Results of the Past and Problems of the Present Plus]. — 2018. — Vol. 7. — № 2 (42). — P. 39-42. [in Russian]
6. Wegberg A.M.J. The Complete European Guidelines on Phenylketonuria: Diagnosis and Treatment / A.M.J. Wegberg, A. MacDonald, K. Ahring [et al.] // *Orphanet J Rare Dis*. — 2017. — Vol. 12. — № 1. — P. 162.
7. Hal'chickij S.E. Issledovanie narushenij metabolizma fenilalanina i ego derivatov pri nasledstvennykh i jekzogennykh giperfenilalaninemiakh [A Study of Metabolic Disorders of Phenylalanine and Its Derivatives in Hereditary and Exogenous Hyperphenylalaninemia]: dis. ... PhD in Biology / S.E. Hal'chickij. — SPb., 2017. — 162 p. [in Russian]
8. Bushueva T.V. Sovremennyy vzgljad na problemu fenilketonurii u detej: diagnostika, klinika, lechenie [Modern View on the Problem of Phenylketonuria in Children: Diagnosis, Clinic, Treatment] / T.V. Bushueva // *Voprosy sovremennoj pediatrii* [Issues of Modern Paediatrics]. — 2010. — № 9 (11). — P. 57-162. [in Russian]
9. Klinicheskie rekomendacii po diagnostike i lecheniju fenilketonurii i narushenij obmena tetragidriopterina [Clinical guidelines for the diagnosis and treatment of phenylketonuria and tetrahydrobiopterin metabolic disorders]. — M.: Akademizdat, 2014. — 70 p. [in Russian]
10. Wood G. Special Low Protein Foods Prescribed in England for PKU Patients: An Analysis of Prescribing Patterns and Cost / G. Wood, Sh. Evans, A. Daly [et al.] // *Nutriens*. — 2021. — Vol. 13. — № 11. — Art. Number 3977.
11. Borovik T.Je. Dietoterapija pri klassicheskoj fenilketonurii: kriterii vybora specializirovannykh produktov bez fenilalanina [Dietary Therapy in Classical Phenylketonuria: Criteria for Selecting Specialized Products without Phenylalanine] / T.Je. Borovik, K.S. Ladodo, T.V. Bushueva [et al.] // *Voprosy sovremennoj pediatrii* [Issues of Modern Paediatrics]. — 2013. — № 12 (5). — P. 40-48. [in Russian]
12. Klitochenko G.V. Sovremennye vozmozhnosti diagnostiki i terapii fenilketonurii [Modern Opportunities of Diagnostics and Therapy of Phenylketonuria] / G.V. Klitochenko, N.V. Maljuzhinskaja, K.V. Stepanenko // *Lekarstvennyj vestnik* [Medicinal Bulletin]. — 2021. — Vol. 15. — № 1 (81). — P. 24-29. [in Russian]
13. Klassicheskaja fenilketonurija i drugie vidy giperfenilalaninemii: klinicheskie rekomendacii [Classical phenylketonuria and other hyperphenylalaninemia: clinical guidelines]. — 2020. — 112 p. [in Russian]
14. Korneva O.A. Issledovanie krivoj vjazkosti presnogo bezglutenovogo testa v zavisimosti ot koncentracii raznykh vidov muki [A Study of the Viscosity Curve of Unleavened Gluten-Free Dough Depending on the Concentration of Different Flour Types] / O.A. Korneva, E.G. Dunec, O.V. Rudenko [et al.] // *Pishhevaja promyshlennost'* [Food Industry]. — 2019. — № 6. — P. 62-65. [in Russian]
15. Bushueva T.V. Specializirovannye produkty lechebnogo pitaniya dlja detej s fenilketonuriej [Specialized Therapeutic Nutrition Products for Children with Phenylketonuria] / T.V. Bushueva [et al.] — M.: Moscow: SMSC Children's Health, 2018. — 128 p. [in Russian]
16. Nikolaeva E.A. Diagnostika i lechenie biopterindeficitnoj giperfenilalaninemii [Diagnosis and Treatment of Biopterindeficiency Hyperphenylalaninemia] / E.A. Nikolaeva, M.I. Jablonskaja, M.N. Harabadze [et al.] // *Rossijskij vestnik perinatologii i pediatrii* [Russian Bulletin of Perinatology and Paediatrics]. — 2015. — № 2. — P. 66-71. [in Russian]
17. Spronsen F. Key European guidelines for the diagnosis and management of patients with phenylketonuria / F. Spronsen, A. Wegberg, K.K. Ahring [et al.] // *The Lancet Diabetes & Endocrinology*. — 2017. — Vol. 5(9).
18. Vockley J. Phenylalanine hydroxylase deficiency: diagnosis and management guideline / J. Vockley, H.C. Andersson, K.M. Antshel [et al.] // *Genet. Med*. — 2014. — Vol. 16. — № 2. — P. 188-200.
19. Semenova N.A. Issledovanie jeffektivnosti produktov suhih specializirovannykh dlja dieticheskogo lechebnogo pitaniya detej, vzroslykh i beremennykh zhenshhin, bol'nykh fenilketonuriej [A Study of the Effectiveness of Specialized Dry Products for Dietary Therapeutic Nutrition for Children, Adults and Pregnant Women with Phenylketonuria] / N.A. Semenova, G.V.

Bajdakova, N.V. Nikitina [et al.] // RMZh. Mat' i ditja [RMJ. Mother and Child]. — 2019. — Vol. 2. — № 4. — P. 355-360. [in Russian]

20. Madzievskaja T. Novye smesi dlja proizvodstva specializirovannyh makaronnyh izdelij [New Mixtures for Production of Specialized Pasta Products] / T. Madzievskaja, T. Shunkevich, A. Belaja // Nauka i innovacii [Science and Innovations]. — 2014. — Vol. 5. — № 135. — P. 42-43. [in Russian]

21. Lovkis Z.V. Razrabotka nizebelkovykh makaronnyh izdelij dlja pitaniya ljudej s narusheniem obmena fenilalanina [Development of Low-Protein Pasta Products for Nutrition of People with Phenylalanine Metabolism Disorder] / Z.V. Lovkis, A.V. Sadovskaja, Ju.S. Usenja [et al.] // Pishhevaja promyshlennost': nauka i tehnologii [Food Industry: Science and Technology]. — 2020. — Vol. 13. — № 3 (49). — P. 6-11. [in Russian]

22. GOST 5900-2014. Izdelija konditerskie. Metody opredelenija vlagi i suhij veshhestv [GOST 5900-2014. Confectionery products. Methods of determination of moisture and dry matter]. — M.: Standartinform, 2019. — 10 p. [in Russian]

23. GOST 5898-87 Izdelija konditerskie. Metody opredelenija kisljotnosti i shhelochnosti [GOST 5898-87 Confectionery products. Methods of determination of acidity and alkalinity]. — M.: Standartinform, 2012. — 26 p. [in Russian]

24. GOST 5897-90 Izdelija konditerskie. Metody opredelenija organolepticheskih pokazatelej kachestva, razmerov, massy netto i sostavnykh chastej [GOST 5897-90 Confectionery products. Methods of determination of organoleptic quality indicators, dimensions, net weight and constituent parts]. — M.: Standartinform, 2012. — 16 p. [in Russian]

25. Avtomatizirovannyj raschet diety. Fenilketonurija [Automated Diet Calculation. Phenylketonuria] // Dieta FKU [FKU Diet]. — URL: http://pku.asteis.net/help_2.html#diet (accessed: 12.09.2023). [in Russian]

26. Specializirovannye produkty lechebnogo pitaniya dlja detej s fenilketonuriej: Metodicheskoe pis'mo [Specialized Therapeutic Foods for Children with Phenylketonuria: methodological letter] / Comp. by A.A. Baranov, T.Je. Borovik, K.S. Ladodo [et al.] — M., 2012. — 84 p. [in Russian]

27. Rylova N.V. Narushenie obmena fenilalanina [Phenylalanine Metabolism Disorder] / N.V. Rylova. — URL: http://www.rusnauka.com/31_NNM_2013/Medecine/5_143828.doc.htm. (accessed: 12.09.2023). [in Russian]

28. MacLeod E. Nutritional Management of Phenylketonuria / E. MacLeod, D.M. Ney // Annals of Nutrition and Metabolism. — 2010. — Vol. 68(2). — P. 58-69.

29. Soderzhanie fenilalanina v osnovnykh produktah pitaniya [Phenylalanine Content in Staple Foods] // Vmeste so vsemi (Forum «Vse o fenilketonurii») [Together with All ("All About Phenylketonuria" Forum)]. — URL: <https://clck.ru/38VTh6> (accessed: 12.09.2023). [in Russian]

30. Soltanizadeh N. Strategies Used in Production of Phenylalanine-Free Foods for PKU Management / N. Soltanizadeh, L. Mirmoghtadaie // Comprehensive Reviews in Food Science and Food Safety. — 2014. — Vol. 13. — № 3. — P. 287-299.

31. Ivanova Z.A. Razrabotka tehnologii hlebobulochnykh izdelij s ispol'zovaniem risovoj muki [Development of Baked Goods Technology with Rice Flour] / Z.A. Ivanova, F.H. Thazeplova, I.B. Shogenova // Problemy razvitija APK regiona [Problems of Development of Agroindustrial Complex of the Region]. — 2018. — № 3 (35). — P. 168-170. [in Russian]

32. Ajrumjan V.Ju. Himicheskij sostav produktov pererabotki zerna risa i kukuruzy dlja povyshennoj pishhevoj i biologicheskoj cennosti hlebobulochnykh izdelij [Chemical Composition of Rice and Maize Grain Processing Products for Increased Nutritional and Biological Value of Bakery Products] / V.Ju. Ajrumjan, N.V. Sokol, E.A. Ol'hovatov // Polzunovskij vestnik [Polzunovskij Bulletin]. — 2020. — № 3. — P. 3-10. [in Russian]