METHODOLOGICAL ASPECTS OF THE USE OF DRY COMPONENTS OF CHICKEN EGGS FOR FEEDING CHILDREN WITH PHENYLKETONURIA

Svetlana T. Bykova1,* Tamara G. Kalinina1, Tatjana V. Bushueva2,3, Tatjana E. Borovik4

1 All-Russian Research Institute for Starch Products — Branch of V. M. Gorbatov Federal Research Center for Food Systems of Russian Academy of Sciences, Kraskovo, Moscow Region, Russia
2 Scientific Center of Children’s Health, Moscow, Russia
3 Research Centre for Medical Genetics, Moscow, Russia
4 I.M. Sechenov First Moscow State Medical University (Sechenov University), Moscow, Russia

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ABSTRACT
Currently, one of the most important tasks facing science and production is the creation of functional product technologies for use in different diets of the population in order to preserve and improve health, as well as reduce the risks and consequences of various diseases, including hereditary ones, such as phenylketonuria (PKU). The All-Russian Research Institute of Starch Products develops technologies for the production of low-protein starch-based products semi-products enriched with functional ingredients and intended for therapeutic nutrition of patients with PKU. As part of the pilot production, the production of these products is organized. Purpose of work: - to justify the possibility of using dry components of chicken eggs (melange, protein, yolk) to enrich low-protein starch products (noodles, vermichel, ‘spider’) intended for feeding children over 3 years old with phenylketonuria; - evaluate organoleptic properties and efficiency of low-protein starch products enriched in hy-pophenylalanine diet of patients with phenylketonuria older than 3 years.

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1. Introduction

In recent years, studies have been carried out confirming the possibility of using dry ingredients of chicken eggs (protein, yolk, melange) to enrich low-protein starch products [1,2,3]. An important factor determining the feasibility of using enriched starch products in the nutrition of children with phenylketonuria is the low content of protein and, accordingly, phenylalanine. It has been established that the inclusion of new ingredients in small quantities in the composition of dry semi-products allows them to be used in low-protein dietary therapeutic diets of patients with PKU, primarily in the nutrition of children.

Two fundamentally new technological techniques have been developed for introducing dry ingredients of chicken eggs into the starch base: the first is the moisture-thermal treatment of a mixture of corn starch and egg products on a roller dryer, the second is the mechanical mixing of the added ingredients and swellable corn starch.

It was found that the physicochemical properties (swelling, solubility, conditional viscosity) of products obtained using various technological methods are close to each other.

A comparison of the two methods of producing new starch mixtures in combination with dry egg products has shown that it is preferable to use a mechanical mixing method because it is simpler and more economical.

2. Materials and methods

The main ingredients for the production of pasta were: corn starch (GOST 32159–2013) and extrusion corn starch of domestic production «Extramil» (TU9187–001–18020121–2001).

To enrich pasta, dry egg yolk (GOST 30363–2013) and Vegeton Children’s Solution (TU9197–033–5869373–06 with changes 1, 2) were used.

According to the results of previous studies [4,5], it has been found that in the development of the formulation of low-protein products, all three components of chicken eggs can be used as a functional additive, but it is preferable to use dry yolk, which is more suitable in quality composition for baby food. Compared to egg mélange and protein, the yolk has a higher energy value and the smallest amount of protein, and therefore phenylalanine. In addition, it surpasses mélange and protein in terms of organoleptic properties, since it has a pronounced taste and smell characteristic of a chicken egg.

Analysis of the chemical composition of new pasta showed that their protein content per 100 g of dry product is less than 1.0%, fat 3.3–3.6%, ash up to 0.14%, carbohydrates no more than 88.0%, energy value ranges from 1614.1–1617.8 kJ (386.14–387.04 kcal).

The results of the analytical evaluation confirmed that the ingredient composition of these products fully meets the requirements for food products of this class (specialized low-protein products) used in the therapeutic and dietary nutrition of patients with phenylketonuria [6,7,8].

Clinical studies of the safety and effectiveness of these low-protein pasta used for the preparation of low-protein dishes were carried out on the basis of the Advisory and Diagnostic Center of the Federal State Research Institute for Children’s Health of the Ministry of Health of the Russian Federation.

Evaluation of the effectiveness of the use of pasta of low-protein starch enriched for dietary therapeutic nutrition for children over 3 years of age: Vermichel, Noodles, Spider was an incomparable, prospective, single-center, uncontrolled study.

Parents (legal representatives) were informed in detail about the goals and tasks of the work, followed by obtaining written informed consent to participate in the study. Children who met the
Children with inherited amino acid metabolism disorders
3 to 18 years old
(classical phenylketonuria)
n=10

Visit № 1 (before receiving the products under investigation)
Signing of informed consent
Clinical examination, evaluation of anthropometric and physical data
Taking Blood for Phenylalanine Test Form
Product Issuance

Telephone contact № 1 (week 2)
-performance of appetite, attitude to food
(for children under 6 years of age, a “eats” “does not eat” assessment,
for children over 6 years old - according to the 5-point system)
-state of skin (presence/absence of rash)
-gastrointestinal state (frequency, stool character)

Telephone Contact № 2 (Week 3)
-performance of appetite, attitude to food
-the state of the skin-gastrointestinal status

Telephone contact № 3 (week 4)
-performance of appetite, attitude to food
-the state of the skin-gastrointestinal status

Visit №2 (5th week against the background of obtaining investigated products)
Clinical examination, evaluation of anthropometric and physical data
Taking Blood for Phenylalanine Test Form

Figure 1. Study Design

inclusion criteria and had no contraindications to prescribing the above low-protein products were randomly selected for follow-up.

Inclusion criteria:
- Children of both sexes between the ages of 3 and 18 years with classical phenylketonuria confirmed at neonatal screening on a hypophenylalanine diet;
- having written consent of parents/legal representatives to participate in the study.

Exclusion criteria:
- acute infectious disease.

The study design is shown in Figure 1.

Under supervision there were 10 patients with phenylketonuria, of which 4 boys and 6 girls, aged 3 years 8 months to 11 years 4 months, the average age of patients was 8 years 4 months ± 2 years 8 months.

The study used a clinical method that included the collection of history data, physical examination with an assessment of growth and body weight, the overall health of the child at the beginning and at the end of the study, an assessment of the tolerability and organoleptic qualities of low-protein products according to the 5 points system, which was carried out using telephone and online contacts. All data obtained were recorded in an individual registration card.

Laboratory research methods included a biochemical (fluorimetric) method for determining the level of phenylalanine in blood serum before prescribing the test products and against the background of their intake 1–2 days before the end of the study.

Statistical processing of the obtained data was carried out using the STATISTICA 13.0 package (StatSoft Inc., USA). Median (Me) values as well as 5th and 95th percentiles were used to describe the test indicators. The differences between the two dependent groups were assessed using a non-parametric Wilcoxon test. The probability of error (p) was considered statistically significant at values < 0.01

3. Results and discussion

In the course of experimental studies, optimal ratios of starch and dry ingredients of chicken eggs were determined, which is very important for maintaining such a quota of protein, which at the output of the production cycle will be the permissible concentration for a low-protein product. So, to prepare a mixture of starch: dry egg white, the content of the latter should be no more than 5%, for the mixture starch: melange — 10%, and starch: yolk — 15%.

With the addition of egg products and an increase in their quota, the content of amino acids and trace elements in the end product also increases compared to the starting starch, while the amount of phenylalanine varies slightly.

It should be noted that the moisture-thermal treatment of the starch protein mixture can be used to produce an artificial egg substitute as a separate semi-product. The food composition obtained by mechanical mixing of swellable starch and dry egg products can be used as an additive for enriching already existing low-protein products, as well as a necessary ingredient for...
baking bread and a filler in various bakery and vegetable dishes (tins, vegetable balls, cutlets, etc.).

In order to increase the nutritional value of low-protein pasta (noodles, vermichel, “spider”) widely used in medicinal nutrition of patients with PKU, dry egg products were introduced into their composition.

3.1. Product Assignment Procedure

After the initial examination and assessment of nutritive status (anthropometric indicators), each child was assigned all 3 types of the studied products as part of the low-protein menu throughout the follow-up. The products were given home for 30 days in the form of semi-finished products at the rate of 1 bag (1.0 kg) of each product for use as an independent dish (pasta, vermichel, noodles) or a component for the preparation of other dishes (soup, stew, casseroles, etc.).

The investigated products were introduced into the diets of children with phenylketonuria in the form of ready-made dishes instead of previously used similar products of other manufacturers in accordance with clinical recommendations for providing medical care to children with PKU [9] and the fundamental principles of hypophenylalanine diet organization [10,11]. The duration of each product intake was 10 days, in order to avoid uniformity of diet, the products alternated throughout the study.

Organoleptic properties (taste, consistency), as well as the appearance of new low-protein mixtures of starch enriched "Vermichel," "Noodles," "Spider," intended for dietary therapeutic nutrition for children over 3 years old, all children rated "good" and "excellent." Most of them (7 out of 10 children) noted appearance of new low-protein mixtures of starch enriched "Vermichel," "Noodles," "Spider," intended for dietary therapeutic nutrition for children over 3 years old, all children rated "good" and "excellent." Most of them (7 out of 10 children) noted appearance of new low-protein mixtures of starch enriched "Vermichel," "Noodles," "Spider," produced using modern technologies enriched with dry yolk and carotene ("Vetoron" — children’s solution) showed high organoleptic qualities and effectiveness in maintaining the stability of the permissible level of phenylalanine in the blood of children with phenylketonuria Use of the above products allows to expand the range of hypophenylalanine diet and increase its nutritional value.

The positive results of the study indicate the possibility of widespread use of new enriched low-protein starch products in the practice of therapeutic nutrition in diseases requiring a low-protein diet.

### Table 1

| Parameters | Vermichel | Noodles | Spider |
|------------|-----------|---------|---------|
| Appearance | 4.9 ± 0.18 | 4.4 ± 0.41 | 4.8 ± 0.32 |
| Consistency| 4.5 ± 0.5 | 4.45 ± 0.45 | 4.45 ± 0.45 |
| Taste      | 5.0      | 5.0     | 5.0     |
| Bodyweight | 26.6 (14.8;51) | 27.55 (15;51) | 0.0001 |
| Height     | 130.5 (103.5;162) | 130.8 (103.5;167) | 0.0001 |
| Phenylalanine in the blood serum (mg%) | 4.45 (2.4;12.5) | 2.65 (2.0;8,7) | 0.0001 |

### Table 2

#### Dynamics of parameters of body weight, growth and content of phenylalanine in blood serum in children with classical phenylketonuria against the background of taking the analysed products

### 4. Conclusion

Thus, the clinical testing of new pasta low-protein products "Vermichel," "Noodles," "Spider," produced using modern technologies enriched with dry yolk and carotene ("Vetoron" — children’s solution) showed high organoleptic qualities and effectiveness in maintaining the stability of the permissible level of phenylalanine in the blood of children with phenylketonuria Use of the above products allows to expand the range of hypophenylalanine diet and increase its nutritional value.

The positive results of the study indicate the possibility of widespread use of new enriched low-protein starch products in the practice of therapeutic nutrition in diseases requiring a low-protein diet.

### REFERENCES

1. Bykova, S.T., Buravlyova, T.N. (2011). Using the dry ingredients to egg dressing low-protein starch. Storage and processing of farm products, 10, 70–72. (In Russian)
2. Bykova, S.T., Buravleva, T.N., Kalinina, T.E., Borovik, T. V. Food composition for the preparation of infant foods. Patent RF, no. 2462050, 2012. (In Russian)
3. Bykova, S.T., Kalinina, T.G. (2018). The use of dry egg products in the diet of children with phenylketonuria. Proceedings of the Scientific Conference "Topical issues of creating functional products of poultry and other branches of the food industry" Moscow Region: VNIIPP, 78–83. (In Russian)
4. Georgieva, O.V., Bykova, S.T., Kalinina, T.G., Varitsev, P. Yu. (2018). Use of specialised products manufactured in Russia for dietetic (therapeutic) nutrition of infants with congenital disorders of protein metabolism. Pediatric Nutrition, 16(4), 73–79. https://doi.org/10.20955/1727–5784–2018–4–73–79 (In Russian)
5. Bushueva, T.V., Borovik, T.E., Roslavtseva, E.A., Semenova, N.N. (2018). New possibilities of diet therapy for patients with phenylketonuria older than one year. Collection of scientific papers “Molecular biological technologies in medical practice". Akademiizdat, Novosibirsk, 174–180. (In Russian)
6. Bykova S.T., Kalinina T. G. (2019). Low-protein pasta for children patients with phenylketonuria. *Food systems*, 2(3), 20–22. https://doi.org/10.21323/2618–9771–2019–2–3–20–22

7. Program for optimizing the nutrition of children aged 1 to 3 years in the Russian Federation. Methodological recommendations. (2019).Moscow: Scientific Center of Children’s Health. — 36 p. ISBN 978–5–6043946–0–1 (In Russian)

8. Borovik, T.E., Ladodo, K.S., Bushueva, T.V., Timofeeva, A.G., Kon’, I. Ya., Kruglik, V.I., Volkova, I.N. (2013). Dietotherapy of Classical Phenylketonuria: Criteria for Choosing Specialized Phenylalanine-Free Products. *Current Pediatrics*, 12(5), 40–48. (In Russian)

9. Phenylketonuria and tetrahydropterin metabolism disorders in children. Clinical recommendations. [Electronic resource: https://www.pediatr-russia.ru/information/klin-rek/deystvuyushchie-klinicheskie-rekomendatsii/%D0%A4%D0%9A%D0%A3%20%D0%BA%D0%BB%D0%B8%D0%BD%18%D0%BD%18%D0%BA%D0%B8_2017.pdf Access date 15.08.2020] (In Russian)

10. Bushueva, T. V., Borovik, T. E., Fisenko, A. P., Baranov, A. A., Kuzenkova, L. M., Zvonkova, N. G., Timofeeva, A. G., Diakonova, E. Yu., Sattarova, V. V. (2018). Specialized medical nutrition products for children with phenylketonuria. Moscow: Scientific Center of Children’s Health. — 128 p. ISBN978–5–900592–8–7 (In Russian)

11. Borovik, T.E., Ladodo, K.S. (2015). Clinical dietetics of children. A guide for physicians. Moscow: MIA.-720 p. ISBN 978–5–9986–0225–2 (In Russian)

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**AUTHOR INFORMATION**

Svetlana T. Bykova — candidate of technical sciences, Scientific Secretary, All-Russian Research Institute for Starch Products — Branch of V. M. Gorbatov Federal Research Center for Food Systems of RAS, 140051, Moscow region, Kraskovo, Nekrasov str., 11.Tel.:+7–495–557–23–85. E-mail: vniik@arrisp.ru

ORCID: https://orcid.org/0000–0002–6483–1319

*corresponding author

Tamara G. Kalinina — candidate of technical sciences, leading researcher, Department of biodegradable polymers, technological tests and experimental production, All-Russian Research Institute for Starch Products — Branch of V. M. Gorbatov Federal Research Center for Food Systems of RAS, 140051, Moscow region, Kraskovo, Nekrasov str., 11. Tel.:+7–495–557–15–00. E-mail: vniik@arrisp.ru

ORCID: https://orcid.org/0000–0002–9751–6315

Tatyana V. Bushueva— doctor of medical sciences, leading researcher, Laboratory of nutrition of a healthy and sick child, Scientific Center of Children’s Health, 119991, Moscow, Lomonosov avenue, 2, Build. 1. Research Centre for Medical Genetics, 115522, Moscow, Moskvorechye str., 1

Tel.: +7–499–132–26–00. E-mail: tbushueva1@yandex.ru

ORCID: https://orcid.org/0000–0001–9893–9291

Tatyana E. Borovik — doctor of medical sciences, professor, Head of the laboratory of nutrition of a healthy and sick child, Scientific Center of Children’s Health, 119991, Moscow, Lomonosov avenue, 2, Build. 1. I. M. Sechenov First Moscow State Medical University (Sechenov University), 119991, Moscow, Trubetskaya str. 8–2.

Tel.: +7–499–132–26–00. E-mail: nutrborovik@mail.ru

ORCID: https://orcid.org/0000–0002–0603–3394

All authors bear responsibility for the work and presented data.

All authors made an equal contribution to the work.

The authors were equally involved in writing the manuscript and bear the equal responsibility for plagiarism.

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