NHS
Barnsley
Clinical Commissioning Group

Guidance for the Appropriate Prescribing for Phenylketonuria (PKU)

Contents

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1. Overview

Phenylketonuria (PKU) is a rare inherited metabolic disorder that affects around 1:10,000 babies in the United Kingdom. Individuals with PKU have an absence or deficiency of an enzyme called phenylalanine hydroxylase (PAH), which converts the amino acid phenylalanine into tyrosine. PKU is diagnosed via the newborn blood-spot screening programme and can be treated with a special low-phenylalanine diet. However, if left untreated phenylalanine can accumulate in the plasma and reach a concentration that can cause irreversible damage to the brain[1].

2. PKU Protein Exchanges

Phenylalanine is an 'essential' amino acid and cannot be synthesised endogenously therefore must be obtained through dietary intake. The treatment for PKU is based around restricting dietary phenylalanine intake to a level that maintains a safe plasma concentration whilst ensuring sufficient phenylalanine provision to meet the body's biological need[2].

Phenylalanine is present in many protein-containing foods. Given these foods require restriction; it is arguably more common for a PKU diet to be referred to as a 'low protein' diet rather than a 'low-phenylalanine' diet. To regulate phenylalanine intake, 'PKU Protein Exchanges' are used whereby one 'PKU Protein Exchange' refers to the amount of weighed food that contains 50mg of phenylalanine (or 1g of protein for those foods/drinks whereby the phenylalanine content is unknown)[3].

The amount of natural protein (protein in regular food and drink) a PKU patient is able to tolerate each day is dependent upon their individual level of PAH activity. Close blood monitoring can identify the degree of dietary restriction necessary but for many PKU patients, 3-6g of protein per day is all that can be tolerated.

Most foods that are associated with the word 'protein' such as meat, eggs, fish, cheese, and milk are prohibited in the PKU diet due to their high phenylalanine contents. Small, weighed amounts of potato, breakfast cereals, and some vegetables are commonly used foods that make up a patient's 'PKU Protein Exchanges'[2].

3. Prescribable Low Protein Foods

The extremely limited number of naturally occurring low phenylalanine foods available means that prescribable low protein foods are essential. These serve to bulk out the diet, ensure daily energy requirements are met and prevent catabolism, which may result in metabolic instability[2]

4. Prescribable Low Protein Milk Replacements

The required quantities of low protein milk replacements (such as Calogen®, Dalia milk®, Duocal powder®, Lattis®, Loprofin drink®, ProZero®, and SnoPro®) needs assessing on an individual basis by the dietitian. Ensure that prescribing reflects the dietitian's most recent advice to the patient.

5. Prescribable Protein Substitutes

An individual with PKU requires a similar amount of total daily protein to that of a non-PKU individual. However, as patients are restricted to a small amount of natural protein a day, prescribable protein substitutes help bridge the sizeable protein deficit by providing all the other amino-acids except phenylalanine. Many protein substitutes also contain vitamins and minerals. However, if they are not already included within the protein substitute, the dietitian can advise on the best vitamin and mineral preparation to prescribe. Commonly prescribed protein substitutes include (but are not limited to) PKU First Spoon®, PKU Lophlex LQ®, PKU Coolers®, PKU Anamix Junior® and PKU Explore®.

6. Recommendations

- Ensure that the patient is being prescribed low protein foods for an ACBS indication i.e. inherited metabolic disorders, renal or liver failure that requires a low protein diet.
- Ensure that the patient has been assessed by a dietitian and ascertain whether it was agreed that they should continue on a low protein diet.
- Ensure that the dietitian reviews the patient's low protein diet every six months.
- Review quantities of low protein foods being prescribed to ensure appropriateness. Prescribing should neither be excessive nor insufficient for the patient's individual needs.
- Ensure prescribing of protein substitutes are in line with recommendations by the patient's dietitian. Review instructions for use and ensure patients take them as prescribed by their dietitian.
- Ensure that quantities and dosages of milk replacements (e.g. Calogen®, Duocal®, Loprofin Drink®, ProZero® and Loprofin Sno-Pro®) are prescribed in line with the recommendation of the patient's dietitian.

There are over 140 ACBS approved, low protein products available on prescription. Rather than detail a recommended maximum quantity of each food item per month; a maximum number of food units per month for all low protein foods (depending on the age of the patient) are given.

7. Maximum Number of Monthly Units for Prescribable Low Protein Foods in PKU[4]

Age of patient with PKU	Maximum number of units to be prescribed each month
4 months to 3 years	20
4 to 6 years	25
7 to 10 years	30
11 to 18 years	50
Adults	50
Pre-Pregnancy/Pregnancy	50

8. Definition of a 'Unit'[4]

ACBS low protein prescribed food	Definition of one unit
Biscuits/biscuit bars	1 pack (200g)
Bread/bread rolls	1 pack (600g)
Breakfast cereals	1 pack/box (400g)
Cakes/breakfast bars/dessert powder mixes /jelly powder/ chocolate substitute spread	1 pack (300g)
Cheese sauce mix	1 pack (225g dry powder)
Crackers/crispbread /mini crackers Croutons/ breadsticks/savoury snacks	1 pack (200g)
Egg replacer/egg white replacer	1 tub/box (500g)
Energy bars (Vitabite or Chocotino)	1 pack
Flour mix/cake mix/waffle mix	1 packet (500g)
Pasta/Rice	1 pack (500g)
Pasta pots / potato pots/pasta in sauce/XPots	1 pack (300g)
Pizza bases	1 box (500g)
Readymade desserts	1 pack (500g)
Sausage/burger mixes/fish substitutes	1 pack/box (360g dry powder)
Soups	1 box (112g dry powder)
Yoghurt substitute	1 tub (400g)

9. Sapropterin (Kuvan®)

There is to be no prescribing of Sapropterin (Kuvan®) in primary care. Sapropterin may be prescribed by the specialist where indicated in line with NICE TA 729.

10. Prescription Charges

Adults with PKU living in England are not medically exempt from prescription charges unless they are entitled to free prescriptions for other reasons[4]. A prescription pre-payment certificate may be beneficial. For more information please speak to your pharmacist or visit https://www.nhsbsa.nhs.uk/help-nhs-prescription-costs/prescription-prepayment-certificates-ppcs

References

1. Van Wegberg. A, M., MacDonald. A, Ahring. K, et al. *The complete European guidelines on phenylketonuria: diagnosis and treatment* (2017). Available at: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5639803/. [Accessed 21st May 21].

2. National Society for Phenylketonuria (NSPKU). *About PKU*. Available from: https://www.nspku.org/. [Accessed 21st May 21].

3. National Society for Phenylketonuria (NSPKU). *New Guide to PKU Food Exchanges.* Available from: https://www.nspku.org/new-guide-to-pku-food-exchanges/. [Accessed 21st May 21].

4. National Society for Phenylketonuria (NSPKU). *Unit Guidelines for the Prescription of Low Protein Foods*. Available from: https://www.nspku.org/documents/?wpdmc=prescriptions. [Accessed 28th May 21].