NHS
Frimley

Medicines Optimisation Position Statement

Position Statement	**Prescribing of low protein products for patients with Phenylketonuria (PKU) in Frimley Health and Care ICS**
Position Statement number:	011
Approved by Medicines Optimisation Group:	October 2022
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Date of planned review	October 2024

Background

Phenylketonuria (PKU) is a rare, inherited, lifelong and potentially serious metabolic disorder. People with PKU cannot break down the amino acid phenylalanine, which then builds up in their blood and brain.
Treatment is a **life-long low phenylalanine (low protein) diet**. This is the only proven treatment that prevents irreversible brain damage, significant delays in development, and hyperactive behaviour with autistic features in children. In adults it prevents neuropsychiatric co-morbidities and maternal PKU syndrome.
Dietary treatment requires:

- severe restriction of natural protein
- supplementation with low or phenylalanine free protein substitute usually supplemented with vitamins, minerals and docosahexaenoic acid (DHA)
- low protein special food

Policy Statement

People with Phenylketonuria (PKU) may be prescribed low protein special foods throughout their lifetime. The amount prescribed can vary dependant on the age of the person.

Prescribing for PKU

There are two aspects to prescribing in a PKU diet:

- PKU supplements (e.g. PKU Anamix Infant/Junior): Recommend add as 'Repeat' as they are required monthly
- Low protein special food options: Recommend add as Acute or Variable Repeat

PKU patients are reliant upon Advisory Committee on Borderline Substances (ACBS) prescribed low protein special foods. These products are not available to purchase in supermarkets. They are important to:

- improve variety in the diet
- help attain acceptable metabolic control and growth by providing essential calories
- help dietary adherence

The amount of low protein products prescribed will vary from patient to patient and should be determined on an individual basis, usually by a **specialist metabolic dietitian** who may be contacted for additional advice. Guidelines are given which are calculated according to energy requirements. Low protein special foods are expected to provide up to 50% of estimated daily energy requirements in patients on a very low protein diet (with up to 10g/day of natural protein).

Protein requirements are calculated on a 'unit basis'. The maximum number of units required per month varies depending on age. Low protein milk replacements are excluded from this monthly unit allocation.

Age of patient with PKU	Recommended maximum number of low protein items to prescribe each month
4 months -3 years	20 units
4-6 years	25 units
7-10 years	30 units
11-18 years	50 units
Adults	50 units
Pre-pregnancy/pregnancy	50 units

Definition of low protein units

ACBS low protein prescribed food	Definition of one unit	Suggested size of unit
Pasta/Rice	1 box	Up to 500g
Pasta pots / potato pots / pasta in sauce Promin X-Pots	1 pack/box	Up to 300g
Flour mix/cake mix/waffle mix	1 pack/box	Up to 500g
Bread/bread rolls	1 pack	Up to 600g
Pizza bases/pizza base mixes	1 pack	Up to 300g
Crackers/crispbread /mini crackers /croutons/ bread sticks/savoury snacks	1 pack/box	Up to 200g
Sausage/burger mixes/fish substitutes	1 pack/box	Up to 360g dry powder
Cheese sauce mix	1 pack	Up to 225g dry powder
Soups	1 box	Up to 112g dry powder
Breakfast cereals	1 box/pack	Up to 400g
Egg replacer/egg white replacer	1 tub/box	Up to 500g
Yoghurt substitute	1 tub	Up to 400g
Biscuits/biscuit bars	1 pack/box	Up to 200g
Cakes/breakfast bars/dessert powder mixes /jelly powder/ chocolate spread substitute	1 pack/box/tub	Up to 300g
Readymade desserts	1 pack	Up to 500g
Energy bars (Vitabite or Chocotino)	1 pack/box	Up to 175g

The above units are intended as a guide only as package size is variable.
Any pack size greater than the suggested weights should be estimated as more than 1 unit.

The British Inherited Metabolic Diseases Group provides detailed lists of low protein milks and low protein special foods, found here: https://www.bimdg.org.uk/site/formularies.asp

To ensure that patients with PKU receive a consistent product supply of the correct low protein foods and protein substitutes, their **specialist metabolic dietitian may register them with a third-party provider.**
The following companies have home delivery agreements
– Vitaflo at Home (Vitaflo and Fate products)
– Homeward (Nutricia Metabolics products)
– Dial a Chemist (Mevalia, Promin, Taranis and Metax products)
– HealthNet (Cambrooke products)
• GPs should identify in the pharmacy text box the name of the third-party provider

See: *Supporting patients with Phenylketonuria (PKU) in the surgery: a guide for GP administrative staff'.* NSPKU_A4_GP_info_leaflet_6.pdf

References: (accessed September 2022)

British Inherited Metabolic Diseases Group: https://www.bimdg.org.uk/site/formularies.asp

NHS Choices: Phenylketonuria - NHS (www.nhs.uk)

National Society for Phenylketonuria (NSPKU UK): www.nspku.org Helpline: 030 3040 1090