

PRESCRIBING POLICY FOR LOW PROTEIN FOODS IN PATIENTS WITH PHENYLKETONURIA (PKU)

The prescribing of low protein essential foods is supported for those with diagnosed Phenylketonuria (PKU).

Prescribing is limited to a set number of units as detailed within this policy.

BACKGROUND

Phenylketonuria (PKU) is a rare but potentially serious inherited disorder.

In people with PKU the enzyme required to break down the amino acid phenylalanine is either missing or greatly reduced. Phenylalanine builds up in the blood and brain leading to brain damage.

PKU is diagnosed during the new-born blood spot screening. The disorder can be treated by a low phenylalanine diet¹.

WHY ARE LOW PROTEIN FOODS PRESCRIBED?

People with PKU can only eat a very limited diet and the normal foods which can be eaten are mainly fruit and vegetables which have a low calorie density. Most nutritious staple foods are not suitable.

Low protein foods are prescribed in order to provide variety to the diet, to provide the calories for growth, and to prevent hunger which in turn may cause the patient to eat higher protein foods. Low protein foods are expected to provide 50% of the daily energy requirements in patients with PKU³.

Ensure that the patient is assessed by a dietitian before being prescribed a low protein diet. The patient's overall diet and the continued need for a low protein diet should then be reviewed by the dietitian every six months.³

RECOMMENDED PRESCRIBING QUANTITIES

Recommended maximum number of units of low protein foods for each age group²

<i>Age of patient with PKU</i>	<i>Recommended maximum number of low protein items to prescribe each month</i>
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 one unit of ACBS low protein prescribable food ²

<i>ACBS low protein prescribable foods</i>	<i>Definition of one unit*</i>
Biscuits/biscuit bars	1 packet (up to 200g)
Bread/ bread rolls	1 pack (up to 600g)
Breakfast cereals	1 box or pack (up to 400g)
Cakes/ breakfast bars/ dessert powder mixes/ jelly powder/ chocolate substitute spread	1 pack (up to 300g)
Cheese sauce mix	1 pack (225g of dry powder)
Crackers/ crispbreads/ mini crackers/ croutons/ bread sticks/ savoury snacks	1 pack (up to 200g)
Egg replacer/ egg white replacer	1 tub/box (up to 500g)
Energy bars (Vitabite or Chocotino)	1 pack
Flour mix/ cake mix/ waffle mix	1 packet (up to 500g)
Pasta/ rice	1 pack (up to 500g)
Pasta pots/ potato pots/ pasta in sauce/ XPots	1 pack (up to 300g)
Pizza bases	1 box (up to 500g)
Ready-made desserts	1 pack (up to 500g)
Sausage/ burger mixes/ fish substitutes	1 pack/box (up to 360g dry powder). One sachet is not the same as 1 box.
Soups	1 box (112g dry powder)
Yoghurt substitute	1 tub (400g)

*Any pack size that is greater than the stated weights should be estimated as more than 1 unit.

These quantities do not include low protein milk replacements or protein supplements.

Low protein foods are covered by the Advisory Committee on Borderline Substances (ACBS) regulations as per part XV of the Drug Tariff. Prescriptions should be marked "ACBS".

Prescribing should be for a maximum of one month at a time (normal CCG policy).

MILK REPLACEMENTS

The required quantities of milk replacements such as Calogen[®], Dalia milk[®], Duocal powder[®], Lattis[®], Loprofin drink[®], Prozero[®], and SnoPro[®] will depend on the patient as recommended by the dietitian. Ensure that prescribing reflects the dietitian's most recent advice to the patient. ^{2,3}

PROTEIN SUBSTITUTES

Protein supplements prescribing quantities should reflect the dietitian's most recent advice to the patient and will vary depending on the patient. Protein substitutes include PKU First Spoon[®], PKU Lophlex LQ[®], PKU Coolers[®], PKU Anamix Junior[®], PKU Explore[®] or glucose polymer supplements. ^{2,3}

SAPROPTERIN (Kuvan®)

There is to be no prescribing of Sapropterin (Kuvan®) in primary care.

PRESCRIPTION CHARGES

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

WHERE CAN PATIENTS GET ADDITIONAL INFORMATION ABOUT LIVING WITH PKU?

The National Society for Phenylketonuria website <http://www.nspku.org/dietary-information/> gives advice and downloadable information about protein exchanges and the quantities of protein in readily available supermarket products.

PATIENT QUERIES OR COMPLAINTS

Any patient queries or complaints should be directed to the Patient Advice & Liaison service (PALS)

- Ipswich & East CCG & West Suffolk CCG: wscg.pals@nhs.net
- North East Essex CCG: 01206 918730 or NEECCG.PALS@nhs.net

REFERENCES

1. NHS Online: <https://www.nhs.uk/conditions/phenylketonuria/>
2. The National Society for Phenylketonuria (NSPKU) Online: <https://www.nspku.org/download/unit-guidelines-for-the-prescription-of-low-protein-foods/>
3. PrescQIPP- 'Appropriate Prescribing for Phenylketonuria (PKU)' Accessed July 2020 online: <https://www.prescqipp.info/umbraco/surface/authorisedmediasurface/index?url=%2fmedia%2f3449%2fb77i-appropriate-prescribing-for-pku-21.pdf>