

Phenylketonuria

Information for GPs about Diet and PKU

PKU is successfully treated by a low phenylalanine diet. Phenylalanine is an amino acid, present in all protein containing foods. The amount of phenylalanine tolerated by each patient with PKU is small, so the dietary treatment for patients with PKU is very strict. All high protein foods are excluded in the diet. These foods include meat, fish, eggs, cheese, nuts, seeds as well as ordinary bread, pasta, flour, biscuits and cakes.

It is now universally recommended that diet is for life (MRC 1993). It is expected that patients with PKU maintain blood phenylalanine levels within strict guidelines. This, therefore, demands lifelong and stringent dietary control.

The low phenylalanine diet consists of 4 parts:

- A daily allowance of phenylalanine is given in the form of 50 mg phenylalanine exchanges (equating to approximately 1g protein) from foods such as potatoes, breakfast cereals, and some vegetables. The amount that is tolerated is only small and usually equivalent to 4-6g/daily natural protein.
- A protein substitute free of phenylalanine, but containing all other amino acids. Many contain vitamins and minerals, +/- carbohydrate and fat.
- Free low phenylalanine foods. These are given in the diet in normal quantities. They mainly consist of fruits, some vegetables, fats and oils and sugar. Low protein special foods e.g. bread, flour and pasta make an important contribution to daily energy intake.
- Vitamins and minerals: only if not added to the protein substitute.

Prescription requirements

Three essential dietary components need to be prescribed from ACBS listed products.

- A phenylalanine-free protein substitute.
- Vitamins and minerals (if not added to the protein substitute).
- Low protein foods which are ACBS listed.

Phenylalanine-free protein substitute

This has several important functions:

- It provides all other amino acids but phenylalanine.
- It supplies a source of tyrosine.
- It suppresses blood phenylalanine concentrations.
- Many contain a source of vitamins and minerals.



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There are a variety of different protein substitutes available in different formats (powders, liquids, tablets, and capsules). The choice of the protein substitute is always tailored round the specific needs of the patient. The patient's Dietitian will have considered all aspects of the diet and will recommend the best protein substitute for the individual with PKU.

Patients must take protein substitute at least 3 times daily. It is essential they do not run out of protein substitute because this will lead to loss of metabolic control. There are two UK companies who produce protein substitutes and both run a 'free' home delivery service (Homeward and Homecare) for protein substitutes.

Name of protein substitute: many of the protein substitutes which are suitable for PKU have similar sounding names to other products which are used for completely different conditions e.g. **PKU Gel** and **PKU Anamix Infant** for **PKU** and **Tyr Gel** and **Tyr Anamix Infant** for **Tyrosinaemia**. To avoid mistakes, *please check the name of the protein substitute before it is prescribed*. It is particularly important to check the exact prefix at the beginning of the protein substitute.

Special vitamins and minerals

If vitamins and minerals are not included within the protein substitute, the Dietitian will advise you on a separate supplement(s) which will supply these nutrients.

Low protein foods

There are several low protein products which have been specifically designed for patients on a low phenylalanine diet and available on ACBS prescription. These include low protein breads, pasta, flour, cake mixes, burger/sausage mixes, dessert mixes, spreads, biscuits and energy bars.

They are essential for 3 important reasons:

- 1. They provide an essential source of calories to support growth and prevent catabolism which may lead to metabolic instability in PKU.**
- 2. They provide bulk in the diet to prevent hunger and consequently help prevent people eating higher protein forbidden foods. The latter would also increase blood phenylalanine concentrations.**
- 3. They provide variety in a limited diet.**

None of the ACBS foods can be considered as luxury items. There are over 100 items available on prescription, and it is important that patients with PKU are allowed some variety and are permitted to use a wide range of these foods. The only way in which they can access these foods is through ACBS prescription; there are few low protein foods which can be purchased from food shops and supermarkets. Patients will request some different low protein items from month to month and this will help them adhere to their diet more easily.

The patient's Dietitian will give guidance on the appropriate number of units of special low protein food to prescribe each month.



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N.B. Many of the low protein foods have similar sounding names to gluten free foods. However, gluten free foods contain protein and could lead to poor blood phenylalanine control if they are prescribed in error. It is important to be aware of this when prescribing low protein food items.

About 40% of the low protein foods are available on a home delivery service (Loprofin products). This is, of course, an optional service but many families do find it helpful. The home delivery service is free of charge. All other products are issued in the usual way through the chemist.

Other important issues:

Aspartame in drugs: Aspartame is an artificial sweetener which contains phenylalanine. It may be added to some drugs and is not permitted in PKU. Please ask the pharmacists to check the label for the addition of aspartame (E951) in all drugs.

Pre-pregnancy/pregnancy: Women if they are planning pregnancy or are pregnant, require particularly stringent dietary control and so need good supplies of all their dietary products. It is essential they are under the care of a metabolic team specialising in PKU management.

Monitoring: all patients with PKU are monitored by regular testing of blood phenylalanine concentrations. Caregivers or patients are usually taught how to take blood samples, collected on a blood card, which are then posted to their hospital. Results are given directly to families, together with guidance on any dietary changes which may be necessary.

Long term follow –up: All patients with PKU should be under the supervision of a hospital team caring for patients with PKU. It is important to check that patients are under the care of a PKU team and have regular contact with their team.

NSPKU website: Further information on low phenylalanine foods that have been ACBS prescribed or any aspect of management of PKU is available from:

www.nspku.org

References

Medical Research Council Working Party on Phenylketonuria. Recommendations on the dietary management of phenylketonuria. *Arch Dis Child*, 1993b, **68** 426-27.