

# Your Patient with PKU

## Information for Pharmacists

- Phenylketonuria (PKU) is an inherited metabolic disorder which, if left untreated results in irreversible brain damage.
- PKU is treated with a strict low protein diet. Natural protein found in food is replaced with a protein substitute and prescribed low protein food products.
- The patient with PKU will require regular prescriptions for amino acid supplements and for low protein foods.
- There are a wide range of protein substitutes available in different forms such as infant 'milks', powders, drinks and gels in different flavours and also tablets and capsules.
- There are over 100 low protein foods available on prescription. Those approved can be found under the borderline substances listed in the drug tariff. A guide to age appropriate quantities is available on the NSPKU website [www.nspku.org](http://www.nspku.org)
- Adults with PKU are subject to the usual prescription charges. It is recommended that adults apply for a prepayment certificate.
- A constant supply of the protein substitute is essential to prevent an increase in blood phenylalanine. There are serious consequences to a disruption in supply of protein substitute.
- The substitute the patient uses will be adjusted periodically, but usually not frequently, it can therefore be ordered in advance to ensure constant availability.
- Low protein products are not available in shops therefore all supplies are obtained by prescription.
- Patients will often prefer an item from one manufacturer over another. Please follow patient preferences for low protein foods to provide variety and encourage adherence to the diet.
- A number of products will need to be ordered as "specials" or directly from the manufacturer.
- As some items will need to be specially ordered your patient will need to order repeat prescriptions in good time.

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### Important

There is a range of protein substitutes available for different metabolic conditions. Some of these products have similar names and packaging. It is critical to check that the products dispensed are suitable for use in the PKU diet. Serious clinical consequences can result if confused.

Similarly many gluten free products are of like appearance and are also not suitable to be used in a low protein diet.

Drugs containing Aspartame should be avoided where possible, unless benefit outweighs risk.

### Emergencies

In an emergency it may be possible to obtain amino acid supplements direct from manufacturers, or the patient's dietitian may have a limited stock.

When new products become available on prescription the manufacturer will be able to tell you if and when they are ACBS approved (allowed on prescription) and how to order them.

### Further help

Your patient's metabolic clinic will be able to discuss any problems or concerns that you have regarding your patient with PKU, or to provide further information and advice.

#### Useful contacts for: *(PATIENT'S NAME)*

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Dietitian Name: \_\_\_\_\_

Dietitian Phone: \_\_\_\_\_

Dietitian Email: \_\_\_\_\_

GP Name: \_\_\_\_\_

GP Phone: \_\_\_\_\_

GP Email: \_\_\_\_\_