PHENYLKETONURIA

(PKU)

Information for GPs on PKU

Phenylketonuria - Dietary Management

PKU is successfully treated by a very low phenylalanine diet. Phenylalanine is an amino acid component of all proteins. This diet is very strict and high protein foods; e.g. meat, fish, eggs, cheese, soya, nuts as well as ordinary bread, biscuits, cakes and pasta are not permitted.

The MRC Working Party report (1) recommended that diet should be continued for life and that blood phenylalanine concentrations should be maintained at even lower levels than previously recommended in the years prior to the 1990's. This demands prolonged and stringent dietary control.

The low phenylalanine diet consists of 3 parts:

1. An allowance of a small measured quantity of phenylalanine given in the form of exchanges from food such as potatoes, baked beans and breakfast cereals. Many patients will only be allowed 4 to 6 exchanges per day. Examples of one exchange are 20g baked beans or 15g cornflakes.

2. The administration of a protein substitute which includes all other amino acids except phenylalanine. Some protein substitutes contain added vitamins and minerals, whereas others consist of non-phenylalanine amino acids only. In the latter group, vitamins and minerals need to be given in addition to the supplement.

3. Free Foods. There are only a few natural foods low enough in phenylalanine to be permitted in normal quantities in the diet. Examples include most fruit, some vegetables, sugar, butter, boiled sweets and some squashes. In addition there are a number of specially produced low protein products such as low protein flour, bread and pasta, which are suitable for PKU and are ACBS listed.

Therefore, three important dietary components need to be prescribed for PKU. They are:

- A phenylalanine free protein substitute
- Vitamins and minerals (if not added to the protein substitute). (see 2)
- Low protein specialised 'foods' which are ACBS listed. (see 3)

Phenylalanine free protein substitute

Adequate quantities of a phenylalanine free protein substitute for all patients on dietary treatment is essential in PKU. Insufficient amounts may result in protein catabolism and higher blood phenylalanine concentrations. There are only a limited number of protein substitutes available. Although vital in the management of PKU, most are not very palatable and are difficult to take.

Each of the protein substitutes has its own merits and drawbacks and the choice and dose of the protein substitute is usually tailored to meet the specific needs and preference of the patient. The patient's dietitian will have considered all aspects of the diet and advise on the best protein substitute to use.

Name of protein substitute

Some of the protein substitutes have similar sounding names, e.g. XP Maxamaid and XP Maxamum or may have similar names to other protein substitutes used in other conditions, e.g. XP Analog for PKU and XPhen,Tyr,Analog for Tyrosinaemia. To avoid mistakes, please check the name of the protein substitute before prescribing and when using products always document the exact prefix at the beginning of the protein substitute.

Special vitamins and minerals

If there are no added vitamins and minerals in the protein substitute, the dietitian will recommend that you prescribe a separate comprehensive vitamin and mineral supplement and advise on the most appropriate one.

Special low protein products

There are a variety of low protein products, e.g. low protein breads, pasta, egg replacer, flour, biscuits and energy bars which have been specifically devised for low protein diets and are ACBS listed for PKU. These provide an essential source of energy as well as adding variety to a very limited diet.

It is important that the patient is prescribed an adequate quantity of these products so that they are able to maintain a good energy intake to avoid protein catabolism.

One of the commonest reasons for poor dietary control relates to inadequate intake of palatable and convenient energy source products. Sometimes patients become bored with one type of biscuit or bread, or achieve better cookery results with one type of flour over another and it is therefore important that they are encouraged to try a wide range of the products available. Hence, they may need to adjust the types and quantities of products on their regular prescription.

It sometimes seems as though some of these foods e.g. Loprofin Chocolate Flavoured Wafers are the equivalent of what would be luxury items for other people. This is not the case and we cannot stress enough how important the prescribing of such foodstuffs is for these patients. The dietitian will provide you with an up to date list of ACBS listed low protein special products.

GPs have some groups of patients (PKU patients could come into this category) who make higher demands on drug budgets. There may be local agreement with the Primary Care Trust to have drug budgets adjusted to reflect this prescribing.

Name of low protein products

Many of the products developed for low protein diets and gluten free diets have similar sounding names and packaging. Errors have occurred with gluten free products being supplied instead of low protein products. Most gluten free products are NOT low in

protein and it is ESSENTIAL that all products are checked before prescribing to ensure they are LOW IN PROTEIN.

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 all drugs prescribed for the presence of aspartame.

Further information on foods passed by ACBS for phenylketonuria and on other aspects of treatment is available on the NSPKU web site.

http:www.nspku.org
For prescription items:
http:www.nspku.org/prescribe.html

Reference 1. Report of the Medical Research Council working party on phenylketonuria. Recommendations on the Dietary Management of Phenylketonuria. Achives of Diseases of Childhood. 1963.**68**. 426-427

Produced by the National Society for Phenylketonuria
and its Medical Advisory Panel.

NSPKU

The National Society for Phenylketonuria (United Kingdom) Ltd.

The Society is a registered charity.
It offers support to PKUs and their families by producing various publications including a quarterly newsletter, organising formal and informal meetings and conferences.

Further information and details can be obtained by contacting:-
The NSPKU Helpline on: 0845-603-9136
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