

The prescription of low protein foods in PKU

People with PKU can only eat a very limited diet. They are unable to eat many nutritious and staple foods: e.g. meat, fish, eggs, cheese, milk, nuts, ordinary bread, pasta and other cereal products. There are few normal foods they can eat in unlimited quantities and these mainly include free fruits and vegetables which have a low calorie density. Therefore, the diet requires supplementation with many low protein foods that are available on ACBS prescription.

This is for 3 important reasons:

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

None of the ACBS foods can be considered as luxury items. There are over 100 items available on prescription, and it is difficult to define how much to prescribe of each item.

In order to give some you some guidance on this, a table is included on page 2 indicating maximum numbers of units of low protein foods to prescribe for different age groups of patients. This has been calculated on energy requirements and in general, low protein special foods are expected to provide between 50% of estimated daily energy requirements in patients with PKU. Some patients eat a variety of different low protein items, but there are many who will eat only a small number of items and eat only low protein bread or crackers as their main energy source. It is therefore not unusual to find that a patient or carer request only flour or biscuits on prescription. Therefore, rather than specify that patients have a maximum quantity of each food item each month; a maximum number of food units per month for all low protein foods (*except milk replacements and glucose polymer supplements*) depending on the age of the patient is recommended, so that patients can have the necessary variety in their diets. The definition of a unit is given below:

ACBS prescribeable foods	Definition of one unit
Pasta	1 box (500g)
Bread/bread rolls	1 packet
Flour mix/cake mix	1 packet (500g)
Breakfast cereals	1 packet
Pasta snack pots	4 snack pots
Pizza bases	2 pizza bases
Sausage/burger mixes	4 sachets
Biscuits	1 packet (150g)
Energy supplements e.g. Duobar, Vitabite	1 packet
Cakes	6 x 40g
Egg replacer	1 packet
Dessert mixes	6 sachets
Low protein spreads	1 tub



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The table does not include milk replacements such as Calogen, Duocal, Loprofin drink, Prozero and SnoPro or glucose polymer supplements. The amount prescribed will vary from patient to patient, so will be determined on an individual basis.

Table indicating recommended maximum number of units of low protein foods for each age group.

Age of patient with PKU	Recommended maximum number of low protein items to prescribe each month
4 months -3 years	15 units
4-6 years	25 units
7-10 years	30 units
11-18 years	50 units
Adults	50 units
Pre-pregnancy/Pregnancy	50 units

NB: this excludes low protein milk replacements: Calogen, Duocal, Loprofin drink, Prozero and SnoPro. It also does not include protein substitutes e.g. PKU Lophlex LQ, PKU Anamix Junior, PKU Gel or glucose polymer supplements.