Guidance for the appropriate prescribing of low protein foods for phenylketonuria (PKU)

Background

Although rare, phenylketonuria (PKU) is the most common inherited metabolic disorder affecting the metabolism of foods containing protein. In people with PKU, the enzyme, phenylalanine hydroxylase, required to convert the amino acid phenylalanine into tyrosine is either absent or deficient. PKU is diagnosed by screening at birth and affects 1 in 10,000 of the population[1].

If untreated, the build-up of phenylalanine in the blood can result in severe brain damage.

Restricting the dietary intake of phenylalanine has been shown to be a successful treatment[1]. Low protein foods are prescribable against the ACBS criteria within the NHS[2]. Prescribing for these patients can be complex and requires regular review to ensure ongoing appropriateness for the patient's dietary needs.

Key recommendations for prescribing for PKU

- Ensure that the patient is being prescribed low protein foods for an ACBS indication i.e. inherited metabolic disorders, renal or liver failure requiring a low protein diet.
- Ensure that the patient has been assessed by a dietitian prior to initiation on a low protein diet
- Ensure that dietitian reviews the patient's low protein diet every six months.
- Review quantities of low protein foods being prescribed to ensure appropriateness based on the maximum number of food units per month, as well as the age of the patient. Prescribing should neither be excessive nor insufficient for the patient's individual needs.
- Ensure prescribing of protein substitutes are in line with recommendations by the patient's dietitian.
- Ensure that changes to a patient's prescribed low protein foods are made only at the request of a dietitian, or in line with NSPKU recommendations.
- Ensure that quantities and dosages of milk replacements (e.g. Calogen®, Duocal®, Loprofin Drink®, ProZero® and Loprofin Sno-Pro®) and glucose polymer supplements are as recommended by the patient's dietitian.
- Ensure that there is no FP10 prescribing of Sapropterin (Kuvan®). This was previously NHSE commissioned but is now no longer commissioned (see below).

The following are included in a PKU diet and, as PKU is an ACBS indication, can be prescribed for PKU patients:

1. Low protein special foods
2. Protein substitutes
3. Vitamins and minerals
4. Milk replacements and glucose polymer supplements

It is recommended that patients have a maximum number of food units per month for all **low protein special foods** depending on the age of the patient.

The definition of a unit is given below[3]:

ACBS food product	One unit
Pasta/rice	500g box
Pasta or potato pots/pasta in sauce	300g
Flour mix/cake mix	1x 500g packet
Bread	600g – 800g
Pizza bases/pizza base mixes	2 pizza bases (300g total)
Crackers/crispbread/ mini-crackers/ croutons/savoury snacks	200g
Sausage/burger mixes	4/2 sachets (125g dry powder)
Cheese sauce mix	1 pack (225g dry powder)
Soups	4 sachets (112g dry powder)
Breakfast cereals/hot breakfasts	375g/6 sachets
Egg replacer/egg white replacer	1 tub
Biscuits	1 packet (150g)
Cakes/breakfast bars/dessert mixes /jelly powder/chocolate spread	240g
Energy bars (Vitabite®)	175g

Recommended maximum number of units of low protein food to prescribe each month[3]

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

Protein substitutes

Protein substitutes should be taken at least 3 times daily with meals in order to keep the phenylalanine levels steady throughout the day, unless otherwise advised by a patient's dietitian.

Review of diet and discontinuation of low protein diet

The European guidelines for the Diagnosis and Management of PKU advises treatment for life for patients with Classical PKU.

Sapropterin

As of April 2019, NHS England no longer routinely commissions sapropterin for the treatment of PKU. This is an updated commissioning position[4]. Available: www.england.nhs.uk/wp-content/uploads/2018/12/Sapropterin-for-Phenylketonuria-all-ages-1.pdf

References

1. PrescQIPP Bulletin 77 (2.1) Appropriate prescribing for phenylketonuria (PKU)
2. British National Formulary Online: https://bnf.nice.org.uk/
3. National Society for Phenylketonuria (NSPKU): The Prescription of Low Protein Foods in PKU, 2017. http://www.nspku.org/sites/default/files/publications/2014%20prescription%20guidelines%20FINAL.pdf Accessed: 19/03/19
4. https://www.england.nhs.uk/wp-content/uploads/2018/12/Sapropterin-for-Phenylketonuria-all-ages-1.pdf

Further information is available from the National Society for Phenylketonuria (NSPKU) www.nspku.org/