Phenylketonuria (PKU) is a rare inherited condition managed with a special low protein diet and daily protein substitute. With preparation, a positive attitude and the right support, children with PKU can participate in all aspects of school life.
What is PKU?
PKU is a rare, metabolic condition inherited from both parents. Children with PKU have a problem with the enzyme that processes phenylalanine (Phe), one of the building blocks of protein. If left unmanaged, high levels of Phe build up in the blood and brain. Over time, this becomes toxic to the brain and causes serious problems with development and learning. However, by following the management plan children develop into normal, healthy adults.

How is PKU diagnosed?
All babies born in the UK and Ireland are screened for PKU at birth by taking a drop of blood from the heel between the 3rd and 5th day of life.

What does the management plan for PKU involve?
- A special, low protein diet
- A protein substitute which must be taken every day

Low protein diet
To prevent high levels of Phe in the blood and brain, the amount in the diet must be limited. As Phe is a building block of protein, children with PKU need to follow a special, low protein diet for life. The amount of protein in the diet is restricted, but not completely taken away: the body cannot make Phe and needs a small amount for essential functions. The amount of protein that a child with PKU can tolerate is monitored by taking regular blood tests; the diet is then altered by a specialist dietitian.

- High protein foods such as meat, chicken, fish, eggs, milk, cheese, nuts, tofu, normal bread and pulses have too much protein to be included in a PKU diet, except in very mild cases where the dietary restriction required is minimal
- Foods containing a lesser amount are then used to carefully measure the amount of protein in the diet. Units called ‘exchanges’ are used

1 exchange = 50mg Phe = 1g protein

- Exchange lists and food labels are used to weigh or measure foods. Foods commonly used include rice, pasta, potato, sweetcorn and crisps – although parents will be able to tell you which foods the child in your class uses and give more information if needed
- Foods very low in protein can then be eaten in normal quantities to provide energy and variety in the diet. They can be split into two groups. The first is those foods naturally low in protein such as fruits and some vegetables, fats, oils, herbs, spices and sugars. The second consists of specially manufactured low-protein foods available on prescription and manufactured by companies such as Nutricia Metabolics e.g. low-protein flour replacement, pasta, milk substitute, cereals, breads and biscuits.

NOTE: If a child with PKU accidentally eats a food not included in their diet, they will not have an immediate reaction. However, you should always inform parents on the same day so that they can adjust the diet appropriately.

Protein substitute
Children need protein for normal growth and development. Because protein in the PKU diet is so restricted, a protein substitute needs to be taken every day at a prescribed dose. This protein substitute contains all of the building blocks of protein with the exception of Phe, and is given in the form of a drink, paste or capsules. Vitamins and minerals must also be included in the diet and are given either as part of the protein substitute or separately. The protein substitute should, where possible, be spread throughout the day and taken at mealtimes alongside protein exchanges.
HANDY TIPS

• It is important that children with PKU only eat foods that are approved by their parents/doctor.

• Parents of children with PKU know best what a child with PKU can and can’t eat.

• Think ahead when organising lessons and events that may involve children in your class eating or drinking: cooking lessons, class parties and school trips are common examples. Notifying parents in advance means that they will be able to help you organise alternative options and avoid the child feeling left out or upset.

• You may think about enforcing a class policy against any student trading food.

• If they have a lunchbox, it is important that the child takes home all uneaten foods from lunch. The parents need to know what was uneaten.

• The more you know about PKU, the more you can help!

Examples of foods from the Loprofin range

Examples of protein substitute drinks

NOTE: Dietary management for PKU varies for each person so the information here is for guidance purposes only. The information provided on this document is in no way intended to replace the care, advice and medical supervision of the child's healthcare professional and/or specialist team.

For more information on PKU please visit the National Society for PKU (NSPKU) website: www.nspku.org or www.lowproteinconnect.com

For more information about protein substitute drinks and the Loprofin range please contact visit www.lowproteinconnect.com or call our metabolic freephone lines: 1800 923 535 (ROI) / 0800 973 216 (NI)