



# PKU and School

## What is PKU?

PKU is a rare inherited condition. PKU stands for phenylketonuria. People with PKU cannot use protein in the usual way. Protein is made up of amino acids, one of which is Phenylalanine or 'Phe' for short. In PKU, the enzyme which breaks down Phe is absent or present in very small amounts. This results in a build up of Phe in the blood. If treatment is not commenced it can result in irreversible brain damage and other health problems.

PKU can be treated completely by a protein restricted diet. \_\_\_\_\_ has been on this diet since birth. With effective treatment children with PKU develop in a normal way. Apart from needing a protein restricted diet \_\_\_\_\_ is exactly the same as other children in the class. He/she is no more likely to suffer from illness than any other child, and can participate in normal school/preschool activities.

PKU is different from an allergy. If a child with PKU consumes food he or she shouldn't eat, there is no immediate reaction. However if this continues to happen on an ongoing basis, there will be a build-up of Phe which causes damaging effects over time.

## How is PKU diagnosed?

All babies in Ireland have a blood sample taken between the 3<sup>rd</sup> and 5<sup>th</sup> day of life, the heel prick test. This is used to screen for PKU as well as other conditions. The incidence of PKU in Ireland is 1 in 4500.

## What Does the Treatment Involve?

At present the treatment for PKU is a special low protein diet plus a synthetic protein supplement. **This diet is for life.** It will prevent the build up of excess Phe in the blood, while at the same time providing enough Phe for growth and development. Blood levels are taken regularly to make sure ideal levels are being achieved.

## Principles of the Diet

- Meat, fish, cheese, eggs, milk, milk products, many vegetarian foods, chocolate, bread, pasta, seeds and nuts are rich in protein and therefore phenylalanine, so they are not allowed.
- Protein is not totally excluded as the child requires a small amount of Phe for growth and development. Foods that contain a small amount of protein such as potato, and cereals are given in small measured quantities so that the blood phenylalanine, which is measured regularly, is kept within safe limits. These measured foods, known as protein *exchanges*, are spread out between the day's meals. The number of exchanges that a child is on changes regularly depending on the blood Phe level.
- Most fruit, vegetables, salads and also sugar, jam, syrups and fats such as butter, fat spreads and cooking oils are protein free. There are also many specially manufactured foods which are low in protein. These include low protein bread, biscuits, chocolate, flour, spaghetti and other pastas and milk substitutes. They are available on prescription for the child with PKU.
- As the high protein foods such as meat, fish, cheese and eggs cannot be eaten, they have to be replaced by a special *synthetic protein drink* from which the phenylalanine has been removed. This is given in the form of a drink or paste. Vitamins and minerals are also included in this drink. The synthetic protein drink should be spread out over the day. It is very important for the child's growth and development and it helps with the good control of blood Phe levels. The drink has a strong taste and smell. *It is important that the teacher has a positive attitude towards the synthetic drink as it is a vital part of the PKU diet.* It may be necessary for \_\_\_\_\_ to take the drink once or twice per day at school. Some children can struggle at times to take their synthetic protein drink. Therefore it is very beneficial if the adults around them including teachers can support them and praise their efforts to take the drink. If the child does not finish their synthetic drink at school please inform parents.

## How should this be Managed at School?

It is important that all those who are involved in the care of the child are informed about the diet. As a teacher you can play a valuable role in helping the child manage their diet whilst at school. Avoid being over sympathetic to the child. This restricted diet is important for their ongoing well being. It is important that exact amounts of protein (exchanges) and the synthetic protein drink are taken each day.

## School Lunches

The child's parents will advise on suitable lunches for school. Many families prefer their child to have a packed lunch.

In order to ensure that the child adheres to his/her protein exchanges accurately, it is important to ensure that:

- Food should not be swapped with other children as this would lead to incorrect protein being taken. You may already have some class rules about food and sharing of food. Having class rules prevents the child from feeling isolated or 'different'.
- Only food provided by parents should be given (or foods from the list that parents have provided that are protein free or low in protein)
- Leftover food needs to be kept and sent home in the lunch box to help parents to keep track of the child's protein intake for the day.
- If the synthetic protein drink is taken to preschool/school it is important to ensure that this is taken.
- If the child has eaten any food that has not been provided by parents or is not allowed or if the child has not taken his/her synthetic protein drink, it is important parents are informed that day so changes can be made to the diet for the rest of the day. This can be written into the child's homework journal for example.
- If the class is learning about food or cooking, recipes may be adapted to low protein versions. Talk to the child's parents as they will be able to help you with ideas or alternatively you can contact the Metabolic Unit on 01 8784317 or email [metabolic.dietitians@cuh.ie](mailto:metabolic.dietitians@cuh.ie).

## Treats and Parties

There are a number of sweets allowed freely in the diet and these can be used for treat days or parties. A piece of fruit also makes a good snack food. The majority of fruit is protein free. If parties or treats are to be given at school it would be advisable to check with the parents and have some suitable foods available for the child with PKU. Families can provide you with a list of suitable treats for parties. **Children with PKU are also not allowed to have foods and drinks which contain the artificial sweetener aspartame or E951/E962** as it contains phenylalanine. Many diet drinks and squashes e.g. contain aspartame as a sweetener. **Double check the labels of all foods and drinks to ensure they do not contain aspartame.**

## Difficulties with the Diet

If a child with PKU eats foods they are not allowed there will not be an immediate effect. Accidental intake of high protein foods will not cause any lasting harm. However regularly eating the wrong foods or having more exchange foods that they are allowed can have a detrimental effect long term on the child's brain. Therefore, incidents where the child eats foods that they are not allowed should not be ignored. They need to be addressed with the child. *Parents should be informed if the child takes a food or drink that he or she cannot have.* If non PKU children eat the low protein foods it will do them no harm. However, this should not be encouraged.

Children with PKU can sometimes experience teasing because the food they eat is different. This should be dealt with in the same way as other teasing or bullying behaviour. This can be discussed within the general context of respect for others and acceptance of differences.

## Educational Achievement

Children with PKU should be treated in the same way as all other children in the class. Early diagnosis and careful management of the condition results in normal healthy children who should aim for the same educational goals as their classmates. Young people with PKU have developmental assessments, as part of their routine care within the National Centre for Inherited Metabolic Disorders. They may also have additional assessments, such as a psychoeducational assessment, should there be any concerns about the child.

If you have any concerns regarding the child's educational achievement, or about issues such as behaviour or attention, talk to the child's parents, who will pass on the concerns to the psychologist working in the Metabolic Unit.

*If you have queries or require more information, contact the Metabolic Unit in Temple Street on 01-8784317 or email [metabolic.dietitians@cuh.ie](mailto:metabolic.dietitians@cuh.ie).*