



# Granny's Guide to

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AND OTHERS INVOLVED IN THE CARE OF THE CHILD WITH

PKU



# Phenylketonuria

To most parents the diagnosis of Phenylketonuria (PKU) is devastating news. They may feel that they will be unable to leave the baby or child with anyone else or go back to work as planned. The information below is to help you understand the diet and help you become confident in dealing with it so that the parents can happily leave the child with you without worrying that the child may be given something that is not suitable.

It is also helpful to learn a little about the diet so that you are aware of the needs of the child, but are able to deal with the diet and not feel overwhelmed and anxious about taking on the responsibility.

## What is PKU?

Phenylketonuria (PKU) is a rare inherited condition which prevents the normal breakdown of protein containing foods. It is due to a deficiency of an enzyme (Phenylalanine Hydroxylase), which breaks down protein foods.



Protein are made up of amino acids. One of these amino acids is called Phenylalanine and would normally be changed into another amino acid – Tyrosine. However, in PKU because the enzyme is not very active, or is missing, the phenylalanine is unable to be changed into tyrosine and will accumulate in the blood. If left untreated the build up of phenylalanine will cause the child to develop learning difficulties.

***Phenylalanine***



***Tyrosine***



We are fortunate that there is a test taken shortly after birth on all babies in the UK, to diagnose this rare condition. The heel prick screening test also tests for some other rare, treatable conditions. If the baby has a positive result to the PKU part of the test then dietary treatment will be started immediately and the child will grow up normally.

The **only** treatment for PKU is a special diet. This diet is low in protein to restrict the amount of phenylalanine consumed.

## The diet

### The diet is divided into 4 main parts:

- High protein foods such as meat, fish, eggs, cheese, and milk contain very high levels of phenylalanine and must be avoided. Because we all need protein for growth and repair of tissues such as muscle and skin, the protein for the PKU diet comes from a special protein substitute "medicine". This contains all the amino acids the body needs except phenylalanine. This "medicine" must be given every day; all of it must be taken and should be spread out evenly over the day. The dietician will have told the parents how much to give and when.

- Some foods such as bread, biscuits and pasta, also contain too much protein for the child with PKU and are therefore replaced by special low protein foods provided free on prescription from the G.P.





● There are many foods which have very little or no protein in them and are “free foods”. These are foods that can be given in normal quantities during the day. These foods are mainly fruit, some vegetables, salads, jams, sugars, syrup and fats such as butter or cooking oils. Children with PKU are encouraged not to have too much sugar and fat for the same reasons as children without PKU.



● The body cannot function properly without the amino acid phenylalanine, but it can only be given in very small amounts to the child with PKU. To achieve this we give a measured amount of phenylalanine every day. These are called **exchanges**. Every child will have a set number of exchanges every day and the number will generally not change even as the child gets older and bigger. The exchanges are provided from foods such as breakfast cereals, potatoes and potato products such as chips and crisps.



## How do we know how many exchanges to give?

The number of exchanges the baby or child needs is estimated on the blood tests that are taken regularly. It may seem cruel to prick the finger or heel regularly, but in fact this is the only way we can tell how much phenylalanine is in the blood. We know that children need to be kept within a strict range so that phenylalanine does not harm the developing brain; therefore, we have to check the blood regularly. The blood test then goes to the laboratory, which analyses the sample and reports the result to the doctor and dietitian. If there is too much phenylalanine in the blood then the exchanges are decreased and if there is too little phenylalanine in the blood then the exchanges may be increased.

The majority of children will not have their exchanges increased or decreased significantly as they get older, as the body only requires a very small amount of phenylalanine to function properly.



## The levels are too high!

There are many reasons why the phenylalanine levels may rise or fall once the child is stabilized on the diet. The most common reason is because too much phenylalanine is taken. This can be because:

- The child has inadvertently taken or been given more exchanges than prescribed. Many drinks contain phenylalanine as a sweetener and can be given in error. (Aspartame contains phenylalanine but all drinks will declare if it is present on the label.
- The child has been unwell. Often many illnesses will put up the levels.
- The child has given in to temptation and helped themselves to "extras".
- The exchanges are not measured correctly.
- The prescription foods have been incorrect. Sometimes foods that are gluten free are given out in error by the chemist or G.P instead of low protein foods.
- The full amount of protein substitute is not taken each day. Over a short time the phenylalanine levels will start to rise.

High levels will show up on the blood tests and should be addressed as soon as possible. If action is not taken and high levels continue for a long time, then the child is at risk of harm to the developing brain. This will affect their abilities at school and eventually in work.

Don't panic if the child has taken something they shouldn't have when you are looking after them. They will not become unwell. However, the parent **must** be told when the child is collected. They can make allowances for this later on and will be able to explain any raised phenylalanine levels. Try to prevent the child getting access to forbidden foods and keep temptation out of their reach.

Sometimes high levels will change the child's behaviour, often making them more naughty, moody, forgetful, tired or emotional. It could also lead to a lack of concentration especially at school. This is often the first indication to parents that the levels have gone up. They can then take steps to investigate what has been taken.



## What can I do to help?

Being a grandparent, child minder or babysitter of the child with PKU can be daunting, the diet is often difficult to learn and there are many things to remember to give throughout the day. It is also hard to deny many of the foods that you want to give such as chocolate, as these are often the treats that most children are given. Sometimes it is hard not to think that a little will not do any harm. However many children with PKU can only tolerate a very small amount of protein and tiny amounts of foods that must be avoided can make a big difference to blood levels. In addition, giving forbidden foods may result in the child developing a liking for these foods and make the diet more difficult for the child to adhere to.

One of the most important factors in managing the diet is being consistent. The majority of the children will want to know why they cannot eat the same foods as you and it is important that you discuss with the parents what answer you will all give. If you say that the high protein foods will make the child sick, and they try them and are not physically sick, then they will not believe you and carry on taking them. Everyone looking after the child must try to have the same answer to provide consistency.

The majority of children will try to take extras when they think they are not seen. You may find sweet papers in pockets, school bags or hidden around the room. The parents and all carers of the child must be consistent with dealing with these "extras". It is no use one person being lenient and the parents being stricter. The child will soon learn who will give them what they want. All extras should be reported to the parents that same day so that they can make adjustments to the diet.

Sometimes there are problems with getting the child to take the protein substitute. This is where grandparents or a child minder can come into their own. Many children will take the substitute for someone else, but not mum. If this is the case then try to work out how you manage and then mum and dad can try the same thing at home.

It may be that the parents are rushing out to work and this can lead to the child getting extra attention by refusing the drink. If this is the case then arrange to give the mix in a more relaxed atmosphere when you are alone and less stressed.

## What can I give the child to eat?

The majority of parents will supply the food for the child to eat during the day when at a child minder's. However, there may be occasions when the food is spoilt for one reason or another, is forgotten or the child is with you longer than expected. The following suggestions may help you to decide what to give in these circumstances. It is a good idea to keep a supply of the favourite foods and a supply of the protein substitute for these times.

Also keep a note of the number of exchanges the child should be given over the day and a note of what you have given, so that the parents do not overdo things once at home. You should check the number of exchanges allowed and the amount of protein substitute given, on a regular basis in case any changes have been made.

The following may help you decide what to give:

### Meals:

Sandwiches made with low protein bread or rolls and butter or suitable margarine filled with -

Jam, salad vegetables, coleslaw, vegetable sandwich spread.

Low protein crackers and butter with any of the above toppings.

Low protein pizza with vegetables and tomato topping

Salad

Low protein soup with low protein bread or rolls.



For dessert the following can be given:

Ready to eat jelly – check it is not sugar free or contains aspartame

Fresh fruit or fruit salad

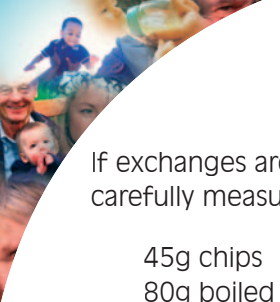
Tinned fruit – best to give fruit in natural juice

Many sorbets do not contain protein – check the label with mum or dad

Sponge puddings or cakes with low protein flour.

Custard made with low protein milk substitute





If exchanges are used then the following count as 1 exchange and must be carefully measured:

45g chips

80g boiled or mashed potato

1 small packet crisps (look on the label and the packet should only contain 1g protein)

55g fried potato shapes

45g boiled rice



## **Drinks:**

The child with PKU will have their protein substitute spread out over the day as a drink and this can make the child thirsty. Plain water is one of the best drinks to give. If you are giving any other drinks then all labels must be checked for Aspartame. This is an artificial sweetener and contains phenylalanine. If drinks containing aspartame are given to the child with PKU then their levels will rise. It is best to ask mum or dad to supply suitable drinks, but if you are out on a trip and need extra drinks then always check the label before giving, even if you have used the drink before. All drinks which contain aspartame will state it on the label and say "contains a source of phenylalanine". Pure fruit juices can be given in small quantities which you would dilute with water. The low protein milk substitutes can be given in limited quantities.



## **Snacks**

Fresh fruit and low protein biscuits can be given freely. Bananas should not be given too often as they have a little more phenylalanine than most fruits. The low protein biscuits and crackers are obtained on prescription from the G.P and you should arrange to keep a supply in the home. These foods can take a few days to obtain from the chemist so give plenty of notice to the parents, when you need more supplies, so that they can be ordered.



## Sweets

There are several sweets that are low in protein and can be given. However, try not to give too many to make up for the diet being so restrictive, as they will harm the teeth and spoil the appetite for meals. There is a list of suitable sweets available on the NSPKU manufacturers lists and ask mum or dad to give you a copy.

The following are the most common sweets to give:

- Low protein high energy bars – supplied on prescription
- Boiled lollies
- Jelly tots
- Polo mints – not the sugar free variety
- Chewits
- Refreshers
- Starburst
- Skittles



Sweets with gelatin are not suitable and labels should be checked for this.

Penny sweets that do not have a list of ingredients are best avoided.

It is helpful to keep a box of treats that can be used when necessary and the treats do not always have to be sweets, you could use stickers or cheap toys or pencils.

The NSPKU have produced an "Pictorial Guide" to the diet. This contains lots of pictures of suitable foods, which can be given in normal quantities. You can help the child personalize the guide by adding in pictures of new free foods.

An order form can be obtained from:  
NSPKU, PO Box 26642, London, N14 4ZF.



## **Don't Worry!**

Looking after the child with PKU can be a bit daunting to start with, but ask the parents for as much information as you need. Do not worry that you will do anything wrong – just check before giving anything you are not sure about first. If the parents mention that the levels of phenylalanine have gone up, do not feel that it is something you have done, there are many reasons and the parents will want to know why.

As mentioned above, the most common reason will be that the child has helped themselves to “extras”, this happens in the home as well as outside. The majority of children will try to take extras at some point; they would not be children if they didn't. The main thing is to find out what, why and deal with it sensitively.

Anyone helping to look after the child with PKU is part of the team that will ensure that the child grows up to be independent, know about their diet and fulfill their potential. By being part of this process and learning about the diet, you will help to reassure the parents that they do not have to give up work, or give up going out because they worry about the diet.

Above all, be positive about the diet. This will help the family accept the condition positively.

### **For more information**

look on the NSPKU website: **[www.nspku.org](http://www.nspku.org)**

**Or**

phone: **0845 603 9136** email: **[info@nspku.org](mailto:info@nspku.org)**

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