



Metabolic.ie

National Centre for Inherited Metabolic Disorders

Sláinte Leanaí Éireann



Children's Health Ireland



A GUIDE FOR THOSE LOOKING AFTER A CHILD ON A LOW PROTEIN DIET

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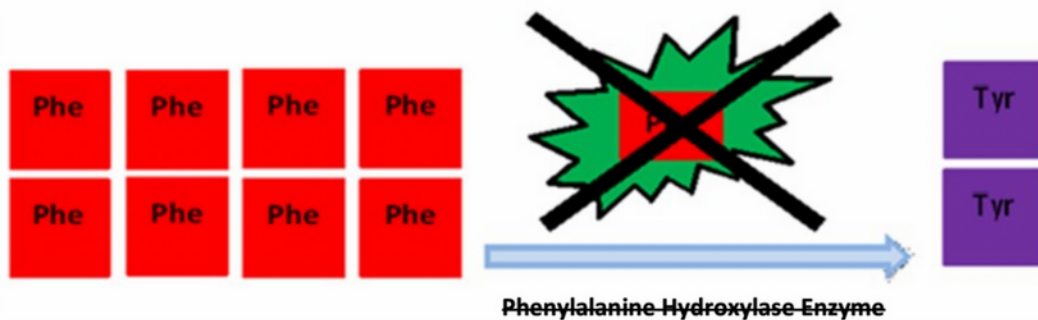
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What is Phenylketonuria?

To some parents, the diagnosis of phenylketonuria (PKU) can be difficult news. They may feel they will be unable to leave their child with anyone else or go back to work as planned. The information in this booklet is to help you understand and become more confident in dealing with the diet required for PKU. This can support the parent to be less worried that their child's diet is managed safely.

PKU is a rare, inherited metabolic disorder, that affects the normal breakdown of protein-containing foods. Protein is one of the main nutrients in our diet. When protein is eaten, it is broken down by enzymes the body makes into things called amino acids. One of these amino acids is phenylalanine (phe). Phe is used to make another amino acid called tyrosine.



In a person without PKU, phe (pronounced fee) is made into tyrosine (TYR) by the body with help from an enzyme called Phenylalanine Hydroxylase (PAH). People with PKU are born without any, or very little amounts of the enzyme PAH. So, phe can't be made into TYR and the can build up in the blood. If left untreated, this can cause damage to the brain, behavioural problems and learning difficulties.

Fortunately, the heel-prick test, taken shortly after all the births of babies in Ireland, screens for PKU. This allows for a diagnosis soon after birth and for immediate treatment with a low protein diet and supplementation. The low protein diet for PKU is a life-long diet and must be adhered to.

Once adhered to, the effects of PKU can be avoided allowing the child to grow and develop normally. They can meet all usual developmental and social milestones. PKU will not hold a child back, they are healthy children with a managed condition and will never have 'blue light' medical emergency due to this condition.



The Diet

The diet is divided into 5 main parts

Foods to avoid

1

High protein foods such as meat, fish, eggs, cheese and milk contain very high levels of phe and must be avoided. Foods that contain a sweetener called aspartame should be avoided as this is a source of phenylalanine. Other foods such as breakfast cereals, potatoes, bread, pasta and rice may only be consumed in measured amounts

Free Foods

2

Some foods naturally have little or no protein and can be eaten freely in the diet. These are foods that can be given in normal quantities during the day. Examples include most fruit, vegetables, salads and small amounts of butter or margarine, jam and honey. All children are encouraged not to have too much sugar or fat.

Low Protein Food substitutes

3

Some foods like bread, pasta, rice and biscuits also contain too much protein for a child with PKU and so are replaced by special low protein foods that are provided free on prescription from the pharmacy.

Synthetic Protein

4

Children need protein for growth and development. To meet this, protein in the diet of a child with PKU comes from synthetic protein. This protein is made by scientists and contains all amino acids except phe, vitamins and minerals.

Natural Protein

5

Children with PKU need a small amount of phe to help them grow. To achieve this, we give a special amount of phe everyday. The amount we give is counted in **exchanges**. An exchange is 1g of protein. Every child will be advised a number of exchanges per day. The exchanges are often provided from foods like breakfast cereals, potatoes, pasta or rice.

1. Be aware of foods to be avoided

2. Be aware of foods that can be eaten freely

3. Low protein substitutes

4 Offer synthetic protein, if required

5. Work with the family around exchanges



Blood Levels, Results and Exchanges

How do you know how many exchanges to give?

The dietitian gets information from blood results that are taken regularly from heel or finger prick blood samples. The sample is measured in the laboratory in Temple Street and the result is reported to the dietitian.

There is a range the phe blood result should be within. If the phe in the blood sample is too high, then the dietitian may recommend lower exchanges. If the phe in the blood sample is too low, the dietitian may recommend exchanges are increased

Range for blood levels

Under 12 years of age = 120 - 360 umol/L

Over 12 years of age = 120 - 600 umol/L



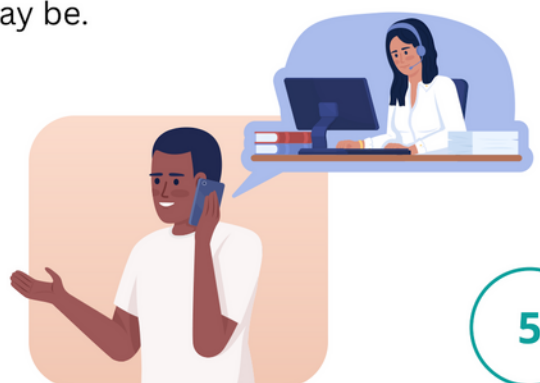
What can cause raised levels?

There are many reasons why phe levels may go up or down even when a child is on the diet. The most common reason for high phe is because too much phe has been consumed for example;

1. The child has accidentally taken or been given more exchanges than advised. This is known as 'over-exchanging'
2. The exchanges are not measured properly
3. Foods or drinks containing aspartame have been consumed
4. The child has been unwell. Many illnesses will naturally put up the levels e.g. tummy bug, flu, teething etc. When a person is unwell, our bodies breakdown protein in order to try and fight off the illness or infection. For children with PKU, this causes a temporary spike in their phe levels which we can't manage as it is outside of our control/not diet-related.
5. Some medications have aspartame in them, be careful to check the labels and find a suitable alternative

Consequences of raised levels

Sometimes lots of high levels for a few weeks change the child's behaviour, often making them more impulsive, upset, forgetful, irritable, tired or emotional. It can be hard to concentrate, especially at school. This is often the first sign to parents that levels have gone up. They can then take steps to look into what the cause may be.



Exchanges

How do I count exchanges?

The majority of parents will give the foods for the child to eat during the day. However, there may be times that you may need to provide the child with exchanges.

An exchange is equivalent to 1 gram of protein. To measure this we provide lots of guidance on what portion of food is counted as one exchange. To help with this, we often talk in scoops. For example ;

80g of potato or 1 and a 1/2 blue scoops = 1 exchange
1 level blue scoop of sweet corn = 1 exchange
1 blue scoop of cooked rice = 1 exchange
1 blue scoop of tinned spaghetti = 1 exchange

Ask parents for
your own blue
scoop!

We provide lots of guides and booklets on food measurements for exchanges on our website metabolic.ie. The parents should also have some resources.

How do I count exchanges from food labels?

Some labels have the protein content per portion already written on the label. You can then use the below table to calculate the number of exchanges

Protein content per portion	Number of Exchanges
0 – 0.3g	Free
0.4g – 0.7g	½
0.8g – 1.2g	1
1.3g – 1.7g	1 ½
1.8g – 2.2g	2

However, if the protein content per portion is not on the label, to calculate the protein content per portion you will need two things

1. Know the amount or weight of the product to be eaten
2. Know the protein content per 100g

Then use the equation

$$\frac{\text{Weight of the product to be eaten} \times \text{protein content per 100g}}{100}$$

Managing Protein Intakes

Being a relative, child minder or babysitter of a child with PKU can be nerve-wrecking. The diet can be difficult to learn and there are many things to remember during the day. It is also hard to say no to many of the foods that you may want to give, such as chocolate, as these can be treats that most children are given.

Sometimes, it is easy to think that a little will not do any harm. But, many children with PKU can only tolerate a very small amount of protein. Also, giving higher protein foods may result in a child wanting more of these foods, making the diet more difficult for the child to stick to.

Tiny amount of protein foods can make a big difference to blood levels and have harmful effects, so be careful with what you give the child!

One of the most important factors in managing the diet is **being consistent**. The majority of children will want to know **why** they cannot eat the same foods as you. It is important you discuss with the parents about the answer you all give. If you say the protein foods will make the child sick, and they try the foods and are not physically sick, they child may not believe it is dangerous and continue to eat the food.

It is important that everyone looking after the child has the same answers relating to PKU so the child gets the same message.

What can I do if the child takes something they are not supposed to?

Don't panic! They will not immediately become unwell. For example, it does not show like an allergic reaction. However, it is important that the parent must be told. The parent can make changes and cut down on exchanges later on that day and will be able to explain any raised phe levels to the dietitian.

Almost all children will 'cheat' on their diet at some point, often more than once! You may find sweet wrappers in pockets, school bags or hidden. The parents and carers must be consistent in dealing with these 'extras'. It is no use one person allowing it and the other being more strict. All extras should be communicated to the parents for them to adjust for.

Agree with the child's parents what answers, terms and approaches to use relating to PKU and the diet

Managing Synthetic Protein Intakes



Sometimes there are problems with getting the child to take their synthetic protein. This is where other family members or a child minder can really help. Many children will take the synthetic protein for someone else, but not for their parent. If this is the case then try to work out how you manage it and then both parents can try the same thing at home.

Some parents might be rushing out to work and this can lead to the child getting extra attention but refusing to drink. If this is the case, then see if you can offer to give the mix in a more relaxed atmosphere when you are alone and less stressed.

It can be useful to look at things through the eyes of the child who is consuming the synthetic drink. Children sometimes explain that everything in their diet feels very controlled and this should be acknowledged. Saying things like “I think you are really amazing to drink your drink. I can see it is hard for you all you have to do every day, you’re great”.

Approach to take when supporting a child to take their drink

While it can be tempting to tell a child “you can’t do X until you drink your drink”, this is not the best approach. It can also be tempting to tell a child that they’ll get sick, or damage their brain if they don’t take their drink. However, these two approaches can feel like a threat of punishment. In the long term it gives the child a negative association with their drink and their condition. A more positive approach is simply stating “first drink your drink, then you can play”. This makes it feel more like the activity is a reward.

“first drink your drink, then you can play”

If a child is saying that their drink tastes ‘yucky’, it is important that you believe them and empathise with them while standing firm that it should be taken. You can say something like “I’m sorry that it tastes yucky. It’s hard for you to drink (hug)”. “You need to take your drink to stay healthy. I’m here with you, let’s get it done”. Rather than try to distract or deny that it is hard, it can be important to let the child know you understand it is hard for them. This can feel validating for the child, and allow them to move on.

“You need to take your drink to stay healthy. I’m here with you, let’s get it done”



What Can I Give the Child to Eat?

The majority of parents will give the food for their child to eat during the day when at a child minder's. However, there may be times when the food is spoilt, food is forgotten or the child is with you longer than expected. The following may help you decide what to give in these circumstances.

Don't feel overwhelmed. First, check the number of exchanges the child is allowed. Then remember, for nearly every food the child can't have, there is an alternative they can have.

If exchanges are required

The metabolic unit have various 'exchange lists' that include measurements of what foods to offer for exchanges. See some popular examples of 1 exchange foods are as below.

- 2 level tbsp Readybrek (8g uncooked)
- 1/2 weetabix (10g dried)
- 1 heaped tbsp* porridge oats (10g uncooked)
- 1 small/'egg-sized' potato (80g cooked)
- 1/2 medium avocado (75g without stone)
- 1 tsp* dried red split lentils (4g uncooked)
- 1 tbsp* kidney beans (12 beans, 15g cooked)
- 1 tbsp* chickpeas (15g chickpeas, 15g cooked)
- 1 tbsp butter beans (17g cooked)
- 2 level tbsp peas (frozen) (20g uncooked)
- 1 tbsp baked beans (10g uncooked)
- Unsalted plain rice/corn/oat cakes**
- Regular penne pasta (8g uncooked**)
- 1 level blue scoop cooked rice/ couscous/ quinoa
- Measured amount natural yoghurt or nut butter



*tsp = teaspoon

*tbsp = tablespoon

** check food labels

Free foods to offer

We have lots of meal ideas on line at metabolic.ie under 'Low Protein Corner'

Main Meals

- Sandwiches made with low protein bread/rolls and a suitable filling
- Low protein crackers with butter or filling
- Low protein rice/couscous/pasta made with
 - roasted/ stir-fried free vegetables
 - AND tomato based free sauce
 - OR low protein cream cheese spread
- Low protein pizza base with tomato purée, vegetables and low protein cheese
- Stuffed peppers with chipped vegetables and low protein rice



Snacks

There are lists of suitable snacks that parents can provide. These come from the pharmacy and can take some time to arrive, so give plenty of notice when you are running short.



Drinks

It is best that the parents supply suitable drinks. Plain water is one of the best drinks to give, special low protein milk can also be given. Pure fruit juices can be provided in small amounts.



Sweets and Treats

There are suitable lists of low protein sweets and treats that the parents can provide or can be found online at metabolic.ie, Try not to give too many as they can fill them up and they will harm the child's teeth.



Check all food labels for **Aspartame**, which is a sweetener containing phenylalanine and is not permitted in the diet.

Aspartame is found in fizzy drinks, cordial, puddings, chewing gums and more. It is also known as E951 and E962.

Keep track of what the child is eating

It may be useful to use a food diary that can be shared between parents and those looking after a child on a low protein diet. This could be shared back and forth at the beginning and end of the day to ensure the child is getting the correct amount of exchanges.

You could use an electronic spreadsheet, texting system or paper and pen. We have included some templates that were kindly shared by members of our parents focus group.

		Offered	Accepted
Snack	Paul's Food		
	Exchanges		
Lunch	Paul's Food		
	Exchanges		
	Synthetic		

Penny's Food Diary

	Monday	Tuesday	Wednesday	Thursday	Friday
Breakfast					
Lunch					
Snack					
Dinner					
Exchanges					
Synthetic Drink					

Penny's Food

	Exchanges
Breakfast	
<i>Synthetic</i>	
Lunch:	
<i>Synthetic</i>	
Snack	
<i>Synthetic</i>	
Dinner:	
Evening Snack	

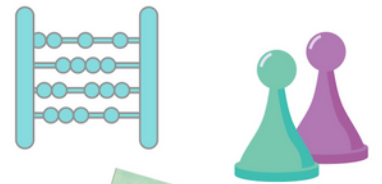
Activities

When looking after a child on a low protein diet, you could consider incorporating some fun low protein food or low protein activities to benefit the child's learning.



You could consider making some **low protein food** as a fun activity. This would help build the child interest in exploring new low protein foods and broaden their food options. We have plenty of **recipes** on our website metabolic.ie. These include recipes for baking foods like low protein bread, bagels and scones. Or, we have savoury recipes to make some tasty dishes using low protein pasta, rice, cous cous or more!

We have lots of **games** as part of our health curriculum that can be shared as an activity to do with a child on a low protein diet. These include games to help the child count exchanges, do calculations and learn all about their special synthetic drink.



We have various animated story books that are specific to different metabolic conditions. Please contact the dietitians and we can send one to you in the post.



Don't Worry!

Looking after a child on a low protein diet can be nerve-racking to start with. It's a whole new way of eating and can take time to learn. Ask the parents for as much information as you need. If the parent mentions that the level of phe has gone up, do not feel it is something you have done, there can be many reasons. Parents will be trying to understand what happened so they can manage it.

As mentioned above, the most common reason will be that the child has helped themselves to 'extras', this can happen in the home as well as outside. Many 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 and why, and deal with it carefully without getting "cross" with the child.

Anyone looking after a child on a low protein diet is part of the team that will help the child grow up to be independent, to know about their diet and to fulfil 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. Remember that what you are doing by helping the child to keep the amount of protein foods low is an act of love, and it will support their brain and body development. It can feel cruel to restrict foods but you are doing it out of love and care.



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