THE PKU HANDBOOK

prepared by the Australasian Society for Inborn Errors of Metabolism (ASIEM)

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Introduction

When phenylketonuria (PKU) is detected, treated early and well controlled, children with PKU have good outcomes. The period between birth and finding out your baby has PKU is too short to cause problems. Early detection and lifelong treatment have been successful in preventing the damage caused by untreated PKU.

This handbook contains information on PKU management, inheritance and phenylalanine (phe) monitoring. It is a tool to assist with the practicalities of a low-protein diet throughout all stages of life and gives details of available support. It also includes information, advice and encouragement from people who live with PKU and their families. The 'Glossary' at the back of the handbook gives simple explanations of all medical terms used throughout the book.

Information in this handbook should only be used in conjunction with your metabolic team and should not replace their advice.

METABOLIC CLINIC CONTACTS

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1

What is PKU?

- Finding out about PKU
- What causes PKU?
- How is PKU detected?
- What is the treatment for PKU?
- The diet for PKU
- Growing up with PKU
- Safe phe levels
- Genetics and PKU: Frequently asked questions
- PKU: The science behind the condition

Finding out about PKU

PKU is short for phenylketonuria, which is a treatable condition where foods containing protein cannot be broken down in the usual way. When PKU is detected and treated early, children are able to reach their full potential.

Finding out your child has PKU happens suddenly and unexpectedly and can be a shock. Most parents have never heard of PKU, so receiving the diagnosis can be frightening and confusing.

What causes PKU?

PKU is an inherited disorder, caused by a faulty gene. Around one in 10,000 babies born in Australia and one in 22,000 babies born in New Zealand has PKU. For a child to inherit PKU, both parents must carry the faulty gene. PKU inheritance is further explained at the end of this chapter.

People with PKU are born with a deficiency of a **liver enzyme** called **phenylalanine hydroxylase (*PAH*)**. *PAH* is needed to process an **amino acid called phenylalanine (phe)**, which is found in foods containing **protein**. Protein is made up of around 20 separate building blocks called amino acids. High-protein foods include dairy products, red meat, chicken, fish, eggs, nuts, beans, tofu and lentils.

Usually, when we eat protein, it is broken down into different amino acids and used for growth, functioning and repair of the body. Because people with PKU can't break down phe in the typical way, it builds up in the blood and damages the brain. People with PKU need phe, but only a *small amount*. Phe intake is explained in more detail in chapter 6.

How is PKU detected?

All babies born in Australia and New Zealand are tested for PKU soon after birth via the Newborn Screening Program, usually on the second or third day. A blood sample is taken from a needle prick on the heel, and the phe level is measured. If it is high, more tests are done to confirm that the baby has PKU.

As soon as the diagnosis is made, the baby is seen by a metabolic clinic team and given a special formula (also called a supplement) to lower the phe to a safe level. Before birth, the mother's blood circulation can process the baby's phe and the level stays within the normal range. Immediately after birth it starts to rise.

When PKU is detected and treated early and well controlled during childhood, PKU will not affect the way children develop. The period between birth and finding out your baby has PKU is too short to cause problems. Early detection and lifelong treatment have been successful in preventing the damage caused by untreated PKU.

What is the treatment for PKU?

Low-protein diet and nutritional supplement

Currently in Australia and New Zealand, PKU is treated with a low-protein diet and a special nutritional supplement for most people.

Regular blood tests to measure the phe level and regular attendance at the metabolic clinic are part of the treatment.

Other potential treatments

1. Medication

For some people with PKU, treatment with a medication called sapropterin (BH4) may be helpful. Refer to chapter 14 on sapropterin for more information.

2. Enzyme replacement therapy

Enzyme replacement therapy uses alternative ways to break down phe, reducing blood phe levels. For example, phe ammonia lyase is given as an injection or tablet. However, this is not yet available in Australia or New Zealand.

Research

Research into future treatments is ongoing around the world. New treatments will be reviewed and included when appropriate.

The diet for PKU

For healthy growth we all need to eat carbohydrate, fat, protein, minerals and vitamins. Phe is found in foods containing protein. People with PKU can't break down phe in the usual way, so the amount of protein in the diet is restricted. A special formula or supplement replaces the protein, minerals and vitamins that would normally be in the diet. The supplement contains all the amino acids except phe. People with PKU obtain the other nutrients they need from food.

'Those first weeks were difficult, trying to work out how much my baby had drunk of her special formula, how much milk, how much was left over and what she needed for the next feed. The personal support from the metabolic team was wonderful.'

Feeding your newborn baby

Newborn babies are given measured amounts of a special PKU formula that contains all the amino acids except phe, and then allowed to breastfeed until they're satisfied. Babies who are not breastfed are given measured amounts of standard infant formula and then special PKU formula until they are satisfied. How much of each formula a baby needs is determined by regular blood testing for phe levels. See chapter 5 'Blood samples'. All babies are different. The precise amount of phe a child with PKU can tolerate varies from child to child. It also varies as the child grows.

The metabolic clinic team – usually a dietitian, doctor, nurse, laboratory scientist and a social worker or psychologist – provides ongoing support.

Feeding your toddler

When solid food is introduced, at around six months, babies with PKU need to eat mainly fruits and vegetables, and some commercial baby foods. As they progress with solids, they can eat a wider range of low-protein foods, including special bread and pasta, and other special low-protein products. Some of these foods can be found in the supermarket; some must be purchased online. The dietitian will help and support you in managing the diet.

The recommended amount of protein from food and supplement is worked out by measuring the blood phe level and making adjustments accordingly.

Growing up with PKU

Maintaining appropriate phe levels in childhood is essential to prevent damage to the developing brain. It is recommended that the treatment be continued for life.

Like any other child, a child with PKU has the potential to grow up and excel in whichever area they choose. Apart from needing a special diet, children with PKU should be treated exactly the same as other children. They're neither more nor less likely to get coughs, colds and other illnesses. Routine immunisations should be given at the usual times and most medicines can be given safely. Check with the metabolic clinic team if you're uncertain.

Without early treatment, features of PKU include:

- babies show signs of slow development by the end of their first year of life
- neurological complications, such as lower intellectual function, hyperactivity or poor coordination
- aggressive behaviour or emotional disturbances
- if left untreated, PKU leads to severe brain damage.

Safe phe levels

Keeping the phenylalanine at just the right level is a balancing act. As long as the level is within treatment range most of the time, your child's development will not be affected by occasional levels that exceed the range.

What is the right phe level?

Metabolic clinics differ on what they consider to be 'acceptable' levels of phe. Occasional higher or lower levels are unlikely to be a problem when overall control is good. General guidelines include the following:

- most clinics agree that for children up to 12 years old, phe levels should be 120–360 micromoles per litre (µmol/L)
- for children over 12 years old, teenagers and adults, an informed decision to accept phe levels above 360umol/L may be appropriate in some cases. However, lower levels are preferable
- women planning a pregnancy or who are pregnant need to have phe levels between 70–250µmol/L to protect the baby (see chapter 13 'PKU and pregnancy')
- be guided by your clinic.

Genetics and PKU: Frequently asked questions

Will other family members be affected?

When a child is diagnosed with PKU, other members of the family may be tested. There is a one in four chance that a full brother or sister of a child with PKU will also have the disorder. Any further babies, including 'half' brothers and sisters, should have a second PKU test even if the first was normal, just to be sure.

How is PKU inherited?

Genes

Our body cells contain instructions, called genes. Genes are important for our body processes. These genes are packaged onto structures called chromosomes. As all of our chromosomes come in pairs, all our genes come in pairs. One copy of each pair is inherited from our mother and the other from our father.

PKU is caused by mutations in both copies of the phenylalanine hydroxylase (*PAH*) gene, which is needed to break down phe in the body. We have two copies of the *PAH* gene, one copy inherited from each parent.

When genes are faulty

A person who has a mutation in one copy of the *PAH* gene but whose other copy is functioning normally is called a 'carrier' of PKU. A carrier is not affected by PKU themselves. Everyone carries some faulty genes.

If both copies of the *PAH* gene have mutations, problems will arise. When two people who are each healthy carriers of a mutation in the *PAH* gene have children, their child may be affected with PKU.

One in 10,000 Australians and one in 22,000 New Zealanders has inherited mutations in both *PAH* genes (one from each parent) and has PKU. Boys and girls have an equal chance of inheriting PKU.

What does having a mutation in this gene mean for the family?

The pattern of PKU inheritance is called 'autosomal recessive inheritance'. As described above, body cells carry two copies of each gene. However, the father's sperm cells and the mother's egg cells carry only one copy. For carriers of a single gene with a mutation, each egg or sperm cell will carry *either* a gene with a mutation *or* a functioning copy of the gene. If by chance both parents carry one copy of the same gene with a mutation, any baby conceived has:

- a one in four chance of inheriting two copies of the gene mutation
- a two in four chance of becoming a carrier like their parents
- a one in four chance of not carrying the gene mutation at all.

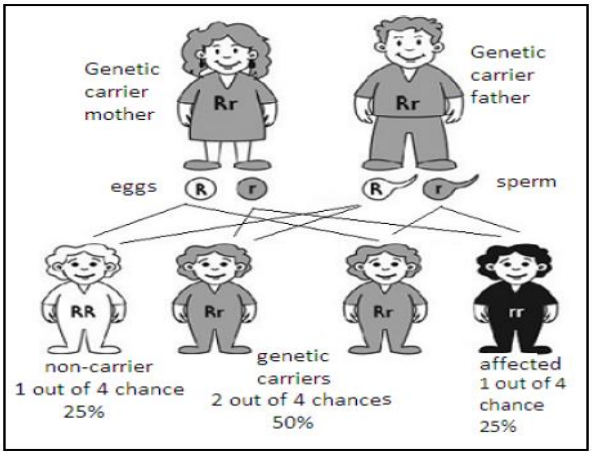


Figure 1: Autosomal Recessive Inheritance (Reproduced with permission from Centre for Genetics Education Autosomal Recessive Inheritance Fact Sheet 7. Available from http://www.genetics.edu.au (Accessed August 2019)

Autosomal recessive inheritance is when both parents are unaffected genetic carriers for the condition. The copy of the gene containing a mutation is represented by 'r'; the correct copy of the gene by 'R'. If both parents carry a mutation in one copy of the PKU gene, there is a one in four chance that their children will have PKU.

If I have PKU, will my children be born with PKU?

If you have PKU and your partner does not, the chance of having a baby with the condition is very low. For this to happen your partner would have to be a carrier. Testing for possible carrier status is available.

In a family where there is already one child with PKU, prenatal testing in a subsequent pregnancy is usually possible. However, because the treatment of PKU is so successful, it is rarely requested. Discuss testing with your PKU doctor or a genetic counsellor.

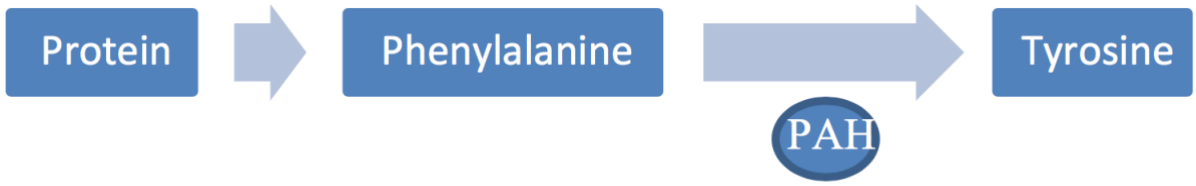
PKU: The science behind the condition

The phe that builds up in PKU comes from protein. All protein comprises an assortment of around 20 chemicals called amino acids. One of these is phe.

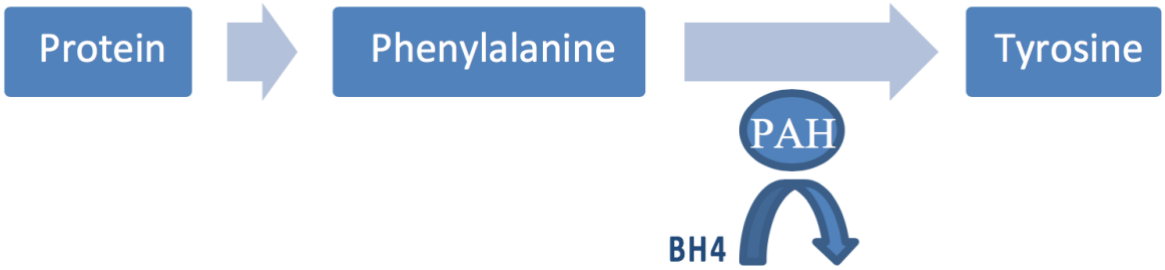
Amino acids in protein are joined to one another, much like beads on a string. Every so often, one of the beads is phe, for example the green beads in the strand below.

When protein is eaten, it is broken up in the stomach into shorter chains of amino acids and then into individual amino acids in the gut. These individual amino acids are absorbed into the bloodstream and processed for use in building muscle, making other chemicals in the body or providing energy.

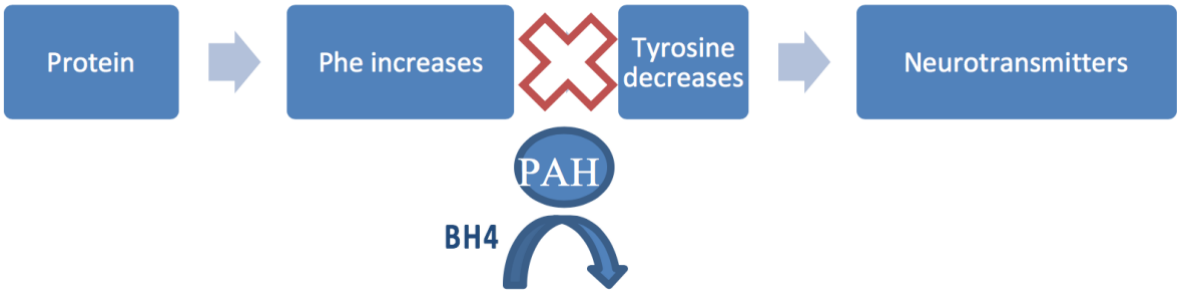
In people who don't have PKU, some of the phe is converted directly into another amino acid, tyrosine, with the aid of the enzyme, phenylalanine hydroxylase (PAH).



In order to do its job, the PAH needs a co-enzyme or helper, called tetrahydrobiopterin or BH4.



People with PKU cannot convert phe to tyrosine. Tyrosine is needed to make the brain's chemical messengers – called 'neurotransmitters'.



Tetrahydrobiopterin (BH4)

BH4 is important too. It is a co-factor for the *PAH* enzyme. Babies found by newborn screening to have a high blood phe level may have investigations to find out exactly where the primary problem is – whether it is with *PAH* or with the BH4 pathway – since the treatment is different.

Some people with PKU may be helped by treatment with BH4 using a medication called sapropterin. Discuss individual eligibility with your metabolic team. Refer to chapter 14 'Sapropterin (BH4)' for more information.

Tyrosine

One use for tyrosine is making the chemical messengers or neurotransmitters in the brain which send nerve signals around the body. It is also needed to make the skin pigment – melanin, and the hormone – thyroxin.

People with PKU cannot make enough tyrosine. For this reason, tyrosine is included in the PKU formula or supplement. Sometimes it is given separately.

Why is the condition called phenylketonuria?

The lack of *PAH* means phe cannot be properly converted for use in the body, so blood levels of phe rise. Eventually, the phe is broken down through alternative pathways and excreted through the urine. The urine contains ketones, one of the breakdown products of amino acids. In PKU, phenyl ketones are excreted, so the condition is called phenylketonuria.

2

What does a diagnosis of PKU mean for my baby and our family?

- Your feelings
- One step at a time
- Sharing the experience
- A team approach
- Explaining PKU

Your feelings

Accepting the diagnosis of PKU can be difficult because your baby looks well. It is alarming to be told that PKU can interfere with your child's brain development, and many parents fear the worst. Words like 'phenylketonuria' and 'phenylalanine' are difficult to understand, and initially it may not be clear what having PKU really means.

'I was really upset. The word "devastating" comes to mind. All I heard was "brain damage". I cried.

Halfway through the interview with the specialist, I thought she could be telling me my baby has something that isn't fixable, but then I realised she was saying we can fix this. After that we just went onward and upward. Sometimes I still feel sad for her but I don't let her see it.'

Once you process the good news that PKU can be controlled, the amount of information about the PKU diet can be overwhelming.

The first few days or weeks can be stressful, especially if you had to wait some time for a full explanation of PKU. It is natural to have feelings of grief, disappointment, sadness or anger about what has happened.

You may also worry about your child's future, and think about the things that will be different and what your child will miss out on. Most parents ask the question, why my child, at some point. Coming to terms with a child's PKU is a grieving process for many parents.

As you learn about the genetic basis of PKU, you may start to feel concerned about your family genes and guilty about passing the condition on. However, you'll also learn that everyone has some faults in their genetic make-up and realise that your child's PKU has come about through the rare chance of two people with the same genetic fault having a child together. It can help to remind yourself that the PKU is not something you could have avoided, and it is not due to anything you have done.

One step at a time

Learning to manage your child's PKU helps in the process of coming to terms with the diagnosis. Most parents begin to feel more positive once they see their child's phe levels come down and start to see how PKU is controlled with the diet. In these early stages, there is often a strong feeling of responsibility, and it is natural to have doubts about how you will cope.

It's normal for your feelings about the condition to change over time. You and your partner/family may deal with the diagnosis of PKU in different ways and feel differently at various stages of the journey. Open communication about how you are coping is vital. Seek support if you need it, or if a family member does. Talking to family, friends, your metabolic clinic team or psychologist can assist.

Sharing the experience

One thing that helps during this time is regular contact with the metabolic clinic team through visits and phone calls. The early weeks and months are also a time to begin sharing the experience with others and allowing trusted family members and friends to support you. Your extended family and friends are always welcome to attend clinic appointments.

Talking with parents of older children with PKU about how they prepare the special foods and what it is like living with the diet can be useful.

Seeing other children with PKU who are growing and developing well is reassuring and can help in developing confidence about the future.

'My first visit to clinic I met a mum of a 9-year-old boy who had PKU. She said, "Think of all the areas of your child's life as books on a large bookshelf. At first it feels like every book is related to PKU. As they grow up you realise that there are so many areas of their life to enjoy – PKU becomes just one tiny book out of the whole library.'

A team approach

PKU is not something parents have to manage on their own. The metabolic team is available to support and guide every step of the PKU journey.

Experience shows it's the children from families that seek medical help, attend clinic and send regular blood samples that do the best.

Explaining PKU

One of the challenges of having a child with PKU is that, because it is rare, few people have heard of it. Finding ways of explaining PKU as simply as possible to yourself, your family, your child and interested others will evolve over time and as the need arises.

Some people will feel ready to tell family and friends about the diagnosis soon after the birth, but others prefer to do so in their own time. PKU is not a visible condition and, apart from the need for a restricted diet, you have a perfectly healthy baby. This allows you to let others know about the condition in your own time. Chapter 9 details specific information for carers, preschool and schoolteachers, doctors and others.

'I realised later that ringing everyone and telling them about PKU and consoling them on the phone made my own acceptance faster. The more people I told and said, look it's all right, it's not that bad, the more I was reassuring myself it was OK.'

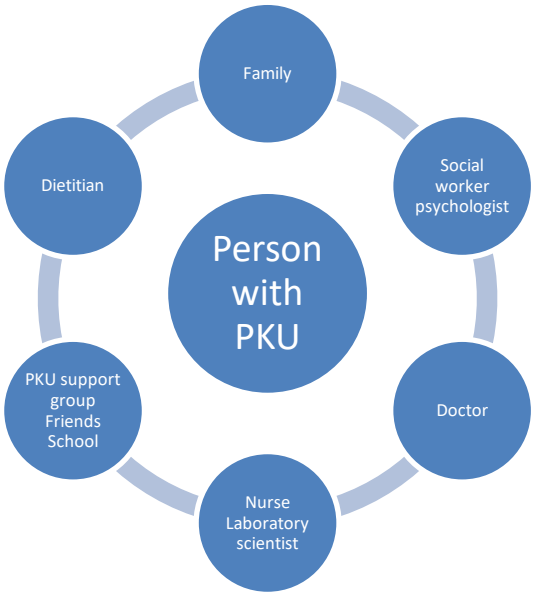


Figure 2: The PKU support circle. You are not alone in managing your child's PKU

Treating PKU: the role of the family

One aspect of having a child with PKU is making a long-term commitment to a treatment plan that will allow your child to reach their full potential.

Although it may seem overwhelming at first, your metabolic clinic team will help you at each stage.

Successful treatment of PKU includes the following for most people:

1. A diet low in phe

The metabolic team regularly determines with you the amount of protein allowed in your child's diet. This is worked out by reviewing their phe levels, age and growth.

2. A daily supplement

This is also calculated specifically for your child's needs. It may be given in the form of a formula, drink, bar, pudding, gel or for some older children and adults, tablets or capsules. The supplement should be taken several times throughout the day to maximise its effect on controlling phe levels.

3. Regular monitoring of blood samples

Blood levels are the most accurate way to measure phe levels in the body. Your clinic will let you know how frequently blood samples are needed. This will vary from twice a week to monthly.

4. Attendance at clinic

When your child is first diagnosed, you will attend clinic frequently for review of growth, feeding, information and support. It is also an opportunity for you to ask questions about your child's PKU. Visits are likely to vary from fortnightly to every three months. As your understanding and confidence grow, the time between clinic visits will increase. Most school-age children (with stable results) visit the clinic every three to six months. Adults may visit annually. In Australia, attending a metabolic clinic is also a requirement for receiving financial assistance from the government. For more information, see chapter 18 'Financial Assistance'.

Individual clinics may provide group education sessions for managing PKU at specific ages. Attending these sessions helps increase your understanding of the condition and gives you the opportunity to connect with other families.

5. Keeping supplies up to date

Part of your responsibility is to make sure you have an up-to-date supply of prescriptions, supplements and low-protein foods, newborn screening cards and blood-taking devices.

'If our child with PKU had been our first instead of our third, or if we had wanted more children after we had her, we would have done so, even knowing the child could have had PKU. Because after three or four years we saw that our daughter's quality of life was no different from that of our other children. She was quite early walking and talking, she was athletic, and she was lovely. She was everything normal, just the same as her siblings.'

3

Feeding your baby

- Breastfeeding
- Formula feeding
- The first few days after diagnosis
- Starting breastfeeds
- Starting standard infant formula
- Phe-free formulas
- How many feeds does your baby need?
- How to express breast milk
- Where to go for help
- Looking after yourself
- What to do if you are unwell
- What to do if you are unable to breastfeed
- How to clean and sterilise feeding equipment
- Stopping breastfeeding

Breastfeeding

Breast milk is usually the ideal food for babies. It contains all the nutrients needed for growth and health. It also helps protect babies from bacterial and viral infections.

You can breastfeed your baby and maintain their blood phe within the target range. However, breast milk alone contains too much phe for babies with PKU. Give your baby a phe-free formula by bottle to take the edge off your baby's appetite before breastfeeding. Most babies quickly learn to cope with the combination of bottle and breastfeeding, or two different formulas.

Formula feeding

If you choose not to, or are unable to breastfeed, standard infant formula is the appropriate substitute, along with a phe-free formula.

The first few days after diagnosis

For the first few days, you may be advised to give your baby just the phe-free formula, and no breast milk or standard infant formula. This will allow the high phe blood level to drop quickly.

The metabolic team will advise you how much phe-free formula your baby is likely to drink. Babies usually make the change to phe-free formula quite easily. If you are breastfeeding, you will need to express to keep up your supply of breast milk (refer to 'How to express breast milk' later in this chapter). When the phe level has come down, you can recommence breastfeeds or standard infant formula feeds.

Starting breastfeeds

Breastfeeds are given ***after*** a measured amount of phe-free formula. If your baby is hungry, offer more breastfeeds after their phe-free formula. Your metabolic team will advise you how to do this. You may think you're making only a small amount of breast milk, but as long as your baby is gaining weight and the blood phe level is in the target range, your baby is getting the amount they need. After the bottle of formula, alternate the breast you offer first. If your baby is not interested in the second breast, don't worry, offer this one first at the next breastfeed.

Starting standard infant formula

Standard infant formula is usually given ***before*** the phe-free formula. If your baby is hungry, offer more phe-free formula. Your metabolic team will guide you how to do this.

The metabolic team will provide a feeding plan for your baby. The order in which you give the phe-free formula and breastfeed or standard infant formula is important but may change from time to time.

Phe-free formulas

Phe-free formulas contain all the nutrients needed for growth, except phe, and come in powdered or ready-made liquid forms. Your metabolic clinic will discuss the available phe-free formulas with you and together you will decide what is best for you and your baby.

The amount of phe-free formula and breast milk or standard infant formula will need to be adjusted regularly to provide the right amount of phe to meet your baby's needs and keep their blood levels in the target range.

Preparing the powdered formula

Refer to the link below for pictorial instructions.

https://raisingchildren.net.au/newborns/breastfeeding-bottle-feeding/bottle-feeding/formula-prep-pictures

- Wash and dry your hands thoroughly before handling bottles and teats and before feeding your baby.
- Sterilise the equipment beforehand (see 'How to clean and sterilise feeding equipment' later in this chapter).
- Measure the formula and water accurately.
- Follow the manufacturer's mixing instructions on the tin, or your dietitian will give you instructions for mixing the PKU infant formula.
- Always use the scoop provided in the formula tin. Level the scoop with a clean dry knife and do not press the powder into the scoop.
- Unless your clinic team advises you to do so, do not add anything to your baby's formula.

Measuring out ready-made formula

The metabolic clinic team will tell you how much and how often to give ready-made phe-free formula. Measure it accurately in the bottle.

Storing the formula

- Prepare each bottle as required or prepare 24 hours' supply at one time.
- Place it in the coldest part of the refrigerator, usually at the back, as soon as it is made – discard any leftover formula after 24 hours.
- Refrigerate sterilised bottles filled with boiled water. Powdered formula is then added at feed time.

At feed time

- Carefully measure the amount of formula required into a feeding bottle.
- Warm the bottle by placing it in a jug of warm water (not boiling). Do not heat the bottle in the microwave as uneven heating can cause hot patches which may burn your baby's mouth.
- Before giving the bottle to your baby, check the temperature by shaking the bottle well and sprinkling a little formula onto the inside of your wrist. It should feel warm, not hot.
- If your baby doesn't drink all of the formula, record what has been drunk and throw out what is left after one hour. Do not reheat used formula.

What to do when you're going out

It is best to take cool boiled water and formula powder or unopened ready-made formula with you to prepare the feed when you need it. A wide-necked vacuum flask of hot water can be used to warm the feeds. Otherwise, take the refrigerated made-up feeds in an insulated bag with an ice pack to keep them cool. Use feeds within two hours, as cool bags do not keep them adequately chilled.

How many feeds does your baby need?

Most babies, whether they're breastfed or bottle-fed, need five or more feeds every 24 hours until they are four to five months old. Also:

- many young babies enjoy extra short snack feeds to settle
- the 'next feed' is considered to be at least an hour after the finish of the previous feed
- your baby may have extra breastfeeds in addition to the phe-free formula and usual breastfeeds
- your dietitian may ask you to keep a record of breastfeeding frequency or infant formula and phe-free formula intake. This helps the team when reviewing blood phe results and giving feeding advice
- provided your baby is well, nothing apart from phe-free formula and breastfeeds/infant formula is needed until around six months of age
- babies should not be given herbal or medicinal teas, or extra vitamins and minerals.

Regular measurements of weight, length and head circumference are taken to make sure your baby is growing well. This can usually be done at your local health centre, and the results will be reviewed by the metabolic clinic team. In NZ, this is completed by your midwife (up to six weeks) then your Plunket nurse.

Talk to the metabolic team if you have any concerns about feeding your baby or feel you need help. Let them know if your baby is unwell or feeding poorly.

How to express breast milk

While your baby is not breastfeeding (during the first two to three days, and any other time breastfeeding is interrupted), you will need to express milk to maintain your supply. You'll need to express six to eight times a day, expressing every three to four hours during the day and less at night.

You can express your milk by hand or with a breast pump (electric or hand pump). Most pharmacies, baby stores and some hospitals sell hand pumps and hire electric pumps. For more information on expressing refer to: https://raisingchildren.net.au/newborns/breastfeeding-bottle-feeding/expressing-working-travelling/expressing-breastmilk

Where to go for help

If you need help with expressing, ask the metabolic clinic team for advice or contact the following:

- the maternal child health nurse (Australia), midwife or Plunket nurse (New Zealand), or lactation consultant
- your health centre nurse
- metabolic clinic nurse
- a midwife
- the Australian Breastfeeding Association, or La Leche League in New Zealand (web address and phone numbers can be found in chapter 19 'PKU Resources').

Explain that your baby has PKU so they will understand why you need advice about expressing milk and/or advice on how to breastfeed.

Looking after yourself

Taking care of yourself is also important.

- Make sure you eat and drink enough to keep up your energy and breast milk supply. Eat healthy snacks if you can't manage full meals.
- Limit tea, coffee and caffeinated drinks to less than three cups a day.
- The safest option when breastfeeding is to avoid alcohol.
- Breastfeeding mothers need to drink an extra three to four cups of fluid a day – water is best for quenching thirst.
- Concern about your baby at this time is natural and this may cause a drop in your milk supply, but it won't affect the quality of your milk.
- Enjoy your baby, rather than focusing on how much breast milk you may be producing – your supply will adjust to your baby's feeding demands.
- Babies with PKU require less breast milk, so a drop in supply is expected.

What to do if you are unwell

Contact your family doctor for advice if you or your baby are unwell, have a temperature, diarrhoea or are vomiting. Prescribed and recommended medicines are generally suitable, but remind the doctor that you are breastfeeding and that your baby has PKU. If your baby is unable to breastfeed, you will need to express to keep your milk supply.

In most cases your baby can and should be put to the breast, even if you are unwell.

If you have a blocked nipple or mastitis, your baby can still breastfeed.

The breast should be drained by your baby suckling or by expressing by hand.

Offer the affected breast first at each feed for a day or two. This empties the breast and will help resolve the problem. If there is no improvement within 12 to 24 hours or you feel unwell, seek medical help. Antibiotics and pain relief may be needed.

What to do if you are unable to breastfeed

Seek advice from your metabolic clinic team as soon as possible. As a guide, your baby can be given unlimited amounts of phe-free formula on demand for up to two days. Offer breastfeeds again as soon as possible.

If you can't resume breastfeeding within 48 hours:

- give expressed breast milk – if available from the freezer – after the usual amount of phe-free formula
- if you don't have breast milk in the freezer, you can give an unlimited amount of phe-free formula for 48 hours, whenever your baby is hungry
- when you are able to obtain some standard infant formula, prepare according to directions on the formula tin and offer it to your baby after the usual amount of phe-free formula
- send a blood test after two or three days so your baby's feeds can be adjusted if needed
- keep in close contact by daily phone calls with your metabolic clinic team until you return to your usual routine. Where possible, it is best to contact the team before switching from breast milk to standard infant formula.

Stopping breastfeeding

In an emergency, follow the guidelines above until you can contact your metabolic clinic team.

When you plan to stop breastfeeding, it is best to discuss this with the metabolic clinic team and do it over two to three weeks.

How to clean and sterilise feeding equipment

The following links provide information and pictorial instructions on cleaning and sterilising feeding equipment:

- https://www.healthed.govt.nz/resource/feeding-your-baby-infant-formula
- https://raisingchildren.net.au/newborns/breastfeeding-bottle-feeding/bottle-feeding/formula-prep-pictures

4

Understanding the PKU diet and protein supplement

- The three parts of the PKU diet
- Different kinds of supplements
- Taking the supplement
- Tips for parents of young children
- When your child refuses the supplement
- Taking the supplement when you're away from home

The three parts of the PKU diet

1. **Formula or supplement**
2. **Counted protein from food**
3. **Uncounted or 'protein-free' food**

People without PKU get all the protein they need from the food they eat. Because protein intake is restricted in the PKU diet, the supplement makes up for what is not provided from food. It helps keep phe levels in the target range, provides all the essential amino acids (excluding or containing only small amounts of phe), tyrosine, vitamins, minerals and trace elements, and aids with fullness throughout the day.

Different kinds of supplements

A variety of supplements are available in Australia and New Zealand. Supplements are made to suit the nutritional needs of people at different ages and are available in various forms to suit different preferences.

The form of protein in the supplement is either:

- amino acid – the protein is cut into single amino acids (see chapter 1 'What is PKU?'). All phe is removed
- glycomacropeptide (GMP) – the protein is cut into small chains of amino acids. These contain only very small amounts of phe and are suitable for some people with PKU.

Supplements are available in a variety of preparations and are suitable for different age groups. Most contain vitamins and minerals, but some do not. Your dietitian will discuss whether additional vitamins and minerals are needed.

'I keep the supplement in a special PKU foods cupboard, and when I have two weeks' supply left, I arrange for the prescription to be filled.'

Supplement preparations

- Powder. Comes in tins or sachets and is mixed into a drink with water, with or without flavouring.
- Gel or thick paste. Eaten with a spoon or taken as a low volume drink, with or without flavouring.
- Liquid. Comes as a single serve, ready to drink.
- Capsules or tablets. A large number is usually required each day and additional vitamin and mineral supplements are needed.
- Bars. Individually portioned.
- Pudding. Eaten with a spoon. Can be consumed frozen.

You can mix and match supplements to suit your lifestyle and preferences.

Supplement supply

The metabolic clinic doctor, nurse practitioner or in New Zealand your PKU dietitian will provide a prescription for the supplement, which is available from pharmacies.

Getting the supplement from the pharmacy

As PKU is a rare condition, your local pharmacist will not have the product in stock. The pharmacist may need to make direct contact with the company that produces the product. Place your order well before you run out to allow for delivery time. It can take a few weeks for a pharmacy to obtain your supplement.

Home delivery

You can also order supplements home delivered, directly from the supplement companies and some hospitals. In Australia, you can contact the company through their website. Ask your dietitian or metabolic clinic team for more information.

Taking the supplement

It is best to take the supplement with meals in several doses spread throughout the day. Also:

- follow the supplement prescription carefully
- if using a ready-made supplement, shake it well. The sediment at the bottom contains important vitamins, minerals and tyrosine
- if your prescription is for scoops, use the scoop provided by the supplement or formula company. Measure level rather than rounded scoops
- if using sachets of formula powder ensure the whole sachet is mixed well with liquid before consumption.

'My child drinks her supplement in a non-spill cup which helps protect the furniture and the carpet. She likes to drink it cold, so I put an ice cube in it.'

Tips for parents of young children

The full amount of prescribed supplement should be taken every day. This is needed for growth, development and to maintain blood phe levels within target range. The supplement:

- is best regarded as part of the meal – you may find it easier to offer the supplement at the beginning of the meal when your child is hungry
- should be given at the same time as 'counted' protein foods
- works best if the volume for the day is divided into three or four small amounts that will not overwhelm your child
- is acidic and could result in damaged teeth if your child sips on it often throughout the day. See chapter 15 'Dental care and PKU'
- offer small amounts from a sipper/straw cup or cup rather than infant formula bottle from nine months of age, so that your infant gets used to drinking it this way.

Right from the beginning, talk to your child about PKU. Explain why the special diet is needed. Treat the supplement as something very special – as a 'magic drink' or 'energy drink' that will make you 'tall and strong'! Give the supplement a special name, such as 'special muscle drink'. Give lots of positive prompts and praise – but let them know taking it is not negotiable.

The supplement may be easier to drink if it is:

- served chilled
- served in a sipper or straw cup to reduce the smell
- flavoured with cordials, fruit flavoured topping, coffee creamers, vanilla, peppermint essence, low-protein milk and flavoured powders or straws.

When your child refuses the supplement

'I know some children refuse their supplement, but mine loves it. It obviously satisfies him, and he comes and asks me for it.'

If your child is having difficulty drinking the prescribed dose of supplement on a regular basis, discuss this with your dietitian. Some reasons may be that:

- your child may be eating or drinking too much at the time they usually drink the supplement
- there is not enough importance attached to having the supplement
- someone may be making negative comments about the supplement
- the supplement could be too much volume for your child to manage or maybe they find it too thick and strong tasting
- your child has noticed how much you want them to drink the supplement and is resisting your efforts
- having the same drink every day has become boring.

Using different flavours, mixing methods and cups can make the supplement more acceptable. Sometimes, changing to an alternative supplement is helpful. Discuss options with your dietitian.

'When our child was young, she was dreadful about taking her supplement and she isn't much better now. I have resorted to bribes, yelling, screaming and crying which was awful. Now I'm trying to be more laid back about it and let her deal with it. She's nearly 15 now and I would say that most of our fighting is about drinking formula. To this day, if I don't ask her if she's had it, she won't go and get it herself.'

Taking the supplement when you're away from home

Tips:

- with young children, take the supplement for your child to drink with a meal. If this is not practical, give some before and after the meal
- use insulated carry bags or a small thermos to keep drinks cool
- encourage your child to take the supplement to school. Many schools are happy to store it in the fridge and remind young children to have it
- some people take extra supplement with them when away from home in case of an unexpected delay.

'My son takes his lunchtime supplement in a pop-top bottle and hands it in at the canteen in an order bag to be sent to the classroom with the lunch orders. This way he can have it cold at lunchtime.'

'I have sachets all the time now. I take three to school and have one during the first period, one after morning break and one just after lunch. I have the last one just before bed. I tried tablets at one stage and they're great because there's no taste at all, but I had to take so many, it wasn't worth it.'

Travelling

You'll find more information about how to manage the diet and supplement when you're away from home in chapter 17 'Overseas Travel'. If you're travelling overseas, you'll need a letter for customs explaining what the supplement is and why it is required. Your metabolic clinic team can provide this letter for you.

5

Blood samples

- Collecting blood samples
- How often are blood samples needed?
- How to take a blood sample (from the heel or finger)
- Troubleshooting
- Tips for success
- Taking a finger prick sample
- Involving children in their blood tests
- Involving children in their blood tests
- Results
- What affects phe levels?

Collecting blood samples

Phe is measured from a small blood sample taken from the heel of babies and toddlers, and from the fingertip as children grow older.

Parents are taught how to collect samples from babies and young children. This usually occurs within the first few weeks. In New Zealand, during the first six weeks after birth, the midwife will assist parents. Grandparents or other carers may also be taught how to take blood. When ready, children will be taught to take their own blood samples.

The procedure is easy to manage once you've had a little practice. You will be supplied with special newborn screening cards (also called blood collection cards) needed for the test. Samples are sent for analysis to the National Testing Centre in Auckland or the Newborn Screening Laboratories in most Australian state capitals.

Blood testing is the only accurate way to measure phe levels in the blood. Tests are taken regularly – your metabolic clinic will let you know how often.

How often are blood samples needed?

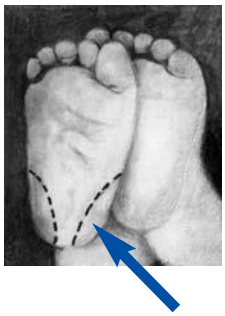
Here is a guide to the frequency:

- during the first few weeks while establishing feeding, blood tests may be needed twice weekly, weekly or as decided by the metabolic clinic team
- once the diet is established, testing can be less frequent, for example, weekly or fortnightly
- after the first 12 months, weekly, fortnightly or monthly tests continue throughout life. Set a reminder in your smartphone, tablet or calendar
- additional blood samples may be needed after an illness or when the levels are too high or too low
- it is best to take the blood sample at the same time of day, when possible, first thing in the morning, before breakfast or three to four hours after a meal and supplement.

How to take a blood sample (from the heel or finger)

This step-by-step guide shows you how to collect a small amount of blood, place it on a card and send it the laboratory for processing (the address may be on the card). At the laboratory, small circles of blood are punched out of the card and tested. A single layer of blood is required for the test to be carried out accurately. For this reason, it is important that the blood completely soak through the card and that blood drops are not placed on top of each other.

1. Fill in the details on the collection card (newborn screening card) using a ballpoint pen (pencil may rub off and felt pen may run). If you don't know the ID number, complete the rest of the card and ask for the number at your next clinic visit. The ID number may be different from the hospital number.
2. You will need:
    - heel or finger pricking device, e.g. a lancet or a diabetic 'pen'
    - blood collection card (newborn screening card)
    - tissues or cotton ball and Band-Aid
3. Wash your hands and make sure the heel or finger has been washed and dried. Use warm water to encourage blood flow.
4. If you're collecting blood from a child, place them in a comfortable but secure position, with one foot or hand free. It is generally easier for one person to hold and comfort the child while a second person holds the foot or hand and takes the blood sample.



5. Select (or have your child select) an area within the recommended sections of the feet (see diagram) to take the blood sample from.
6. Press 'pen' or other device firmly against the heel or finger and push down until released.
7. Wipe away initial drop of blood.
8. Allow a large drop of blood to form on the heel or finger without squeezing.
9. Allow this to drop onto the circle on the card. The drop of blood must be large enough to soak through to the back of the card. *Do not place blood drops on top of each other.* The circles on the card are a guide to the size of the blood drop that is needed.
10. Apply pressure to the puncture site with tissue or cotton wool.
11. Cover the puncture site with a Band-Aid if necessary.
12. Both sides of the blood spot need to dry in the air for at least four hours. For Australian cards, lie the card flat over the end of a bench, or have it sticking out from between the pages of a book, which is lying flat, so both sides of the blood spot can dry. For the New Zealand card, see the back of the card for instructions on how to bend it so both sides of the blood spot are exposed to the air.
13. When it is fully dry, wrap the card in plain paper or for the New Zealand card, place the cover over it.
14. Place in an envelope and send to your Newborn Screening Centre. For babies and young children, it is recommended to use express post or priority post in Australia.

Troubleshooting

Common problems with samples – generally meaning that the laboratory is unable to determine an accurate phe level – include the following:

- blood spot is too small
- using multiple blood spots to fill a circle (i.e. putting one drop on top of another)
- blood smeared over the circle
- blood has not soaked through to the back of the card
- card has got wet
- card has not been allowed to dry slowly in room air
- card has been placed in a plastic bag or container while still wet.

'When I'm taking blood samples, I put warm water in a basin and let my baby kick with pants rolled up and bare feet. It warms up the feet and encourages blood flow. Children love getting their feet wet and it means having the blood test is associated with something fun.'

Tips for success

The following may also help:

- make sure the hand or foot is warm before taking the sample – by putting on socks or gloves, swinging the arm, soaking in warm water or gently rubbing the area
- use gravity to help blood flow by letting the hand or foot hang as low as possible
- have your child in a firm position so they can't wiggle and pull away from the person taking the blood sample
- if blood flow is slow, try gently massaging the calf or lower arm to encourage blood flow.

Taking a finger prick sample

Until a child is about a year old, blood samples are taken from the heel. When children start walking, the skin on their heels becomes tougher and fingertip samples are taken. The procedure is basically the same as for a heel prick sample.

For fingertip samples, a different device may be used. Some clinics provide blood sampling equipment.

Otherwise you will need to purchase a finger pricking device, e.g. a diabetic 'pen', available from pharmacies.

The best ones have a dial that controls how far the skin is penetrated. It is usually recommended that you start with a middle setting and dial up – for deeper penetration, or dial down in numbers – for shallower penetration, depending on how easily the finger bleeds.

While the sample may be taken from any finger, it is best taken from the middle and fourth fingers. Generally, the sides of the fingertip are better to use than the pad. Rotate the finger used to collect the sample.

Involving children in their blood tests

Even relatively young children can play an active role in collecting their blood samples.

- Give them accurate information about what is going to happen – instead of saying 'it won't hurt', let them know that there will be a sharp prick, then when the blood comes it will be put onto the card.
- Let toddlers decide which finger to use for the blood sample.
- Tell them you are going to do it on the count of three, and get them to count with you so they know when to expect it.
- Encourage them to see what a good blood sample they have produced – count the blood drops onto the card.
- Give lots of positive feedback about how well they have done.
- Avoid reinforcing any negative messages, particularly when one parent is doing the blood samples. Phrases such as "isn't Daddy naughty for making your finger hurt" may confuse your child into thinking that the blood sample is taken because they are naughty.
- As soon as the child is old enough to write their name, encourage them to help you fill out their details on the card (if it isn't clear, write it underneath yourself). Involving the child in the process helps make it an achievement rather than an imposition.
- With most of the diabetic pens and lancets, even young children can help press the release button. You can encourage them to press your finger to release the button.
- Be wary of bribing your children over blood tests, especially with food. If you need a reward system, establish a star chart that encourages short, medium and long-term goals.
- Children respond well to routine. Try to collect the blood sample in a consistent way so the child knows what to expect, as fear of the unknown is very real in children.
- Talking in a calm, encouraging voice and distracting your child can help reduce anxiety.

Blood tests are a fact of life for children with PKU. They can't control whether or not the tests occur, but they can control some aspects of the testing. The patterns you establish when they are young will set the foundation for how they cope with blood samples for the rest of their life.

Where are blood samples sent?

New Zealand

The tests are sent to the laboratory at Auckland City Hospital. Your metabolic clinic team will give you pre-addressed and prepaid courier packs for this. Peel off and keep the tracking number in case there are problems with transit to the laboratory.

Australia

Newborn screening services in Australia are provided by five centralised screening laboratories. These are in Western Australia; South Australia – that also covers Tasmania and part of Northern Territory; Victoria; New South Wales – that also covers the Australian Capital Territory; and Queensland – covering part of the Northern Territory.

Results

Blood results are sent to the metabolic team and/or the regional dietitian and paediatrician, depending on where you live. Parents will be contacted if dietary changes are needed after the blood test results are known. In some centres, results are also sent to people with PKU.

What affects phe levels?

It is normal for phe levels to vary a little throughout the day. That is why there is a target range of phe levels rather than one number. Levels are generally higher in the morning because, after an overnight fast, the body starts to break down some of its own stores of protein. During the day, levels fall just after taking the supplement and then rise to a peak before the next supplement is taken.

The following factors cause blood phe to rise above the target range:

- too much protein from food has been consumed
- a child's rate of growth has slowed, and they may be having more protein from food than they need
- not eating enough food or not getting enough supplement for growth and exercise. In this situation the body breaks down its own tissues (e.g. muscle), releasing this into the blood and causing the levels to rise
- sickness, when it may be difficult to eat or take the supplement
- weight loss – the body breaks down protein as well as fat. It is best to lose weight gradually (see 'Controlling your weight' in chapter 10).

The following factors cause blood phe to fall below the target range:

- not eating enough protein from food will result in low phe levels for a short time, then levels will rise as the body breaks down its own protein to get the phe it needs for growth and other processes
- very rapid weight gain (adult) or growth (child) where the body uses phe to build new tissues.

If your or your child's levels are outside the target range, your metabolic clinic team will discuss ways to improve them.

6

Phenylalanine in the PKU diet

- The PKU diet
- How is phe intake counted?
- How to count protein
- Artificial sweeteners
- Medications

The PKU diet

This chapter explains the typical diet for PKU. If you are on alternative PKU therapies such as BH4, refer to chapter 14.

The three parts of the low-protein diet are:

- PKU formula or supplement (see chapter 4 'Understanding the PKU diet and protein supplement')
- counted protein from food (see below)
- low-protein foods (see chapter 10 'The PKU diet')

Protein, including phe, is needed for growth and functioning of the body. People with PKU get most of the protein they need from the formula or supplement. Protein, including phe, is essential for growth and functioning of the body. The table below shows the difference between protein from the supplement and protein from food in the PKU diet.

Protein source	Contains phe that needs to be counted	Provides most of the protein and nutrients in the PKU diet	As your child gets older
PKU supplement	X*	✓	Protein provided increases
Counted protein from food	✓	X	Depends on phe levels

* GMP products contain negligible phe

The table below compares a PKU and non-PKU diet:

Non-PKU diet	PKU diet
• fruit and vegetables • breads, rice, pasta, cereals • meat, chicken, fish, legumes, tofu, eggs, nuts • dairy milks, yoghurt, cheese • fats and sugars	• fruit and vegetables • cereal based foods – e.g. breakfast cereals, crackers and biscuits • low-protein foods – e.g. special flour, bread, rice and pasta • PKU supplement • fats and sugars

Foods such as red meat, chicken, fish, eggs, milk, yoghurt, cheese, nuts, tofu and legumes (e.g. lentils, chickpeas, kidney beans) are usually too high in protein to include in a PKU diet. Some people who have a high protein tolerance may include small amounts of these foods after discussion with their dietitian.

How is phe intake counted?

Foods such as cereals and some vegetables and fruits contain small amounts of protein (and therefore phe). These foods have to be counted in the diet.

How to count protein

Steps for protein counting

For foods without a nutrition information panel on the label, such as fresh fruit and vegetables:

Refer to appendix 1 'Protein counted and free vegetables and fruit in PKU'

For foods with a nutrition information panel on the label:

Refer to appendix 2 'Label reading'

These show the protein amounts in foods, allowing you to work out how much of the daily protein quota you (or your child) will eat at each meal. Your dietitian will discuss protein counting with you.

There are a number of methods used to count protein. Some measure the protein in 'units' (1 unit = 15mg phe), others measure it in 'exchanges' (1 exchange = 50mg phe).

In Australia and New Zealand, protein is counted in 'grams' (1g protein = 50mg phe). For example:

- 1/4 cup green peas = 2 grams
- 1 cup hot potato chips = 3 grams

Artificial sweeteners

Some artificial sweeteners used in foods contain phe and should be avoided. To find out whether a product contains these artificial sweeteners, check the following:

- the ingredient list on the food label
- additives 951 and 962 (also called NutraSweet, Equal, Canderal and aspartame-acesulphame) contain aspartame, which is a source of phe.
- look for a warning statement saying that the product 'contains phe'. These sweeteners are used mainly in diet products such as diet drinks or yoghurts, sugar-free lollies and chewing gum. See example on following page.

Other artificial sweeteners are acceptable in the PKU diet as they do not contain aspartame. Refer to the table below for a list of acceptable artificial sweeteners:

Acceptable	Not Acceptable
950 Acesulphame	951 Aspartame
952 Cyclamate	962 Acesulphame-Aspartame
953 Isomalt	NutraSweet
954 Saccharin	Equal
955 Sucralose	Canderal
956 Alitame	
957 Thaumatin	
961 Neotame	
965 Maltitol	
966 Lactitol	
967 Xylitol	
968 Erythritol	
Mannitol, Sorbitol	

How to check food labels for artificial sweeteners

Below is an example of how to read a food label when checking for artificial sweeteners.

The drink below contains little protein, but it does contain phe (from the sweetener 951, aspartame) so it should be avoided by people on a PKU diet. While a diet product can have little or no protein, and aspartame isn't a protein, aspartame contains a lot of phe.

Ingredients: Carbonated water, colour (150), food acids (330), sweeteners (951), flavour, preservative (211). *Contains phenylalanine.*

Nutrition information		
Servings per package: 3 Serving size: 200 mL		
	Quantity per Serving	Quantity per 100 g (or 100 ml)
Energy	4 kJ 1 Cal	2 kJ 0.5 Cal
Protein	0.1 g	0.05 g
Fat, total	0 g	0 g
- saturated	0 g	0 g
Carbohydrate	0.15 g	0.07 g
- sugars	0 g	0 g
Sodium	20 mg	10 mg

Medications

Sweeteners such as aspartame and aspartame-acesulphame are also used in some medications. Ask your doctor to prescribe medications without these sweeteners. In some situations, it is not possible to prescribe an alternative.

If your child has been prescribed a medication with added aspartame or aspartame-acesulphame, it is recommended that you advise your metabolic clinic.

When buying over-the-counter medications, always try to buy those that do not contain aspartame or aspartame-acesulphame.

Some medications contain a gelatin coating which might also contain small amounts of protein. If the dose of medication is large, it can add to the total amount of protein consumed and should be counted. Check with your clinic or pharmacist about alternative medications without gelatin or protein.

7

Feeding your child

- When to start solid foods
- Introducing solids
- Progressing textures and self-feeding
- How to give solid foods to your baby
- Make eating safe and reduce the risk of choking
- Feeding your child from one year onwards
- Feeding problems
- 7 tips for reducing tension at mealtime
- Resources

When to start solid foods

This usually happens at around six months of age. Babies develop at different rates and some may be ready for solids from four months onwards, but not before this time. It generally takes babies several months to learn to eat solid foods as they get used to new tastes and textures.

Signs your baby is ready to start solid foods

- They are able to sit, with support, and hold their head up.
- They are starting to show signs of being interested in what others are eating.
- They may seem hungry after a full feed, or feed more often than previously needed.

Your metabolic clinic dietitian will help you decide when and how to introduce solids. During this time, it is important to stay in close contact with the dietitian, as it is different from feeding a baby without PKU.

Before you give solids

- Always wash your hands before preparing food and use clean utensils such as bowls or spoons.
- When you make food in advance, do not keep it warm for 'later'. Put it in the fridge or freezer and warm it at the time of the meal.
- Use prepared food within one to two days of cooking.
- Do not reuse a meal that has been partially eaten.
- Mark foods to be frozen with the date packaged or to be eaten by.
- Small quantities of food can be frozen in ice cube trays and thawed as needed.
- Commercial baby foods can be readily used in the PKU diet. Use the nutrition label to calculate protein content. Babies need variety, so use home prepared foods when you can.

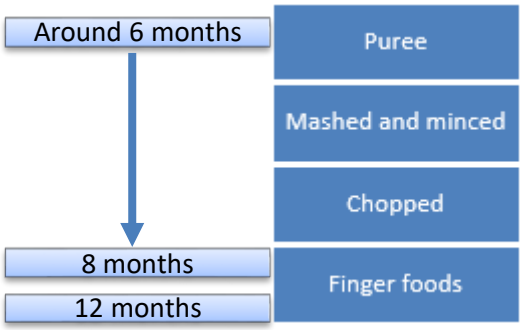
Introducing solids

A traditional way

A traditional way to introduce solids is to start with pureed foods, building to mashed/lumpy foods and then finger foods. Once your baby is able to manage a smooth texture, progress to the next stage, so that they are chewing most normal texture food by one year old.

Babies progress with solids at different speeds. Some may prefer finger foods early, whereas others may need time to master lumpy foods.

Below is a guide to solids progression at different ages using the traditional method.



Baby-led weaning

Baby-led weaning is another way to introduce solids. This allows babies to feed themselves finger foods without spoonfeeding or purees.

The baby sits with the family at mealtimes and joins in when ready, feeding themselves first with fingers and later with cutlery. Discuss this with your dietitian and refer to the link below for more information:

http://www.rapleyweaning.com/assets/blwleaflet2.pdf

Solids can be introduced using a combination of the traditional way and baby-led weaning.

What foods to offer first

- First foods may be smooth, mashed or soft finger foods.
- Uncounted fruits and vegetables are offered first, such as carrots, green beans, peaches, pears, apples and apricots.
- Your baby will enjoy the natural tastes of foods so there is no need to add any fat, sugar, salt or spice to their food.
- Avoid honey until your child is older than one year of age.
- Start with a small amount, such as a teaspoonful or two. There is no need to be concerned if your baby isn't interested. Think of it as a taste at this stage. Gradually work up to larger amounts.
- Offer new foods frequently. Getting used to different tastes is key to increasing the variety of accepted foods. There is no need to wait a few days before trying something new.

Progressing textures and self-feeding

- During the first few weeks and months of introducing solids, babies are usually more open to new textures.
- Babies start to make chewing movements, whether or not they have teeth. It's important to introduce lumpy/finger foods so they develop chewing and self-feeding skills.
- At first, they may push the food out with their tongue. This doesn't mean they don't like it – just that it is a new experience or taste. Eating skills improve quickly over a week or two.
- It can take several offerings before a new food is accepted.
- As your child progresses, try adding the herbs and spices used in family food for taste familiarity. There is no need to add salt or sugar.
- Allow your baby to decide how much to eat. Losing interest, spitting out or pushing the food away, turning their head and closing their mouth are signs they are full. Do not try to force in the last few mouthfuls.

Ideas to progress texture

Mashed and minced	Finger foods
Vegetables mashed with a fork Peeled soft fruits Low-protein pasta/rice or low-protein cheese mixed with purees Lumpy baby cereals Mashed fruit with coconut yoghurt	Hard dry toast made from low-protein bread Peeled soft fruits Strips or pieces of cooked vegetables Low-protein pasta pieces Low-protein crackers

How to give solid foods to your baby

Where and when

- Choose a quiet place and time of day when you and your baby are relaxed. Initially, this will be after a breast or formula feed. When quantities increase, food can be given first.
- Seat your baby in a secure position facing you, such as sitting in a highchair or at a low table.
- Learning to eat is messy! It's normal for food to be dropped and spilt. Placing a plastic mat on the floor makes spills easier to clean up.

Spoon and pouch feeding

- Use a small spoon with smooth edges. Place a small amount of food on the spoon and hold it to your baby's mouth. Press the spoon gently down on baby's lips. When their mouth opens, put the tip of the spoon just inside.
- When feeding your baby from a bowl use two spoons, one for them to practise with and one for you.
- Limit squeezing food from a pouch directly into your baby's mouth. They will learn the skills to eat by feeding from a spoon.
- Do not add solids to your baby's bottle.
- Some babies choose to start with finger foods and skip spoonfeeding altogether.

Introducing a cup

- Offer sips of water from a sipper or straw cup from six months of age.
- Initially, keep giving formula from a bottle and try tiny amounts from the cup.
- Aim to give most fluids from a cup by 12 months old.

Include your baby in family mealtimes from an early age. They will want to have foods they can't eat. You will need to show them which foods they can have. This will encourage good eating habits and help your child to learn that their diet is special. The clinic team will provide tips on how to avoid or manage mealtime problems with siblings and extended family.

Make eating safe and reduce the risk of choking

Avoid the following:

- foods that can break off into hard pieces such as apple pieces
- foods such as raw carrot and celery sticks. Hard foods should be grated, cooked or mashed
- small round foods that can get stuck in children's throats (e.g. hard lollies, grapes, berries, raisins/sultanas, peas, watermelon seeds, popcorn)
- foods with skins or leaves that are difficult to chew (e.g. lettuce, nectarine unpeeled)
- fibrous or stringy foods that are difficult for children to chew (e.g. celery, raw pineapple)

Do the following:

- stay with young children and watch them while they are eating
- have them sit quietly while eating
- never force young children to eat.

Count protein

Your dietitian will meet with you to discuss low-protein foods and protein counting. Initially, foods are weighed and the amount of protein your child eats is added up (see chapter 6 'Phenylalanine in the PKU diet' and appendix 1 'Protein counted and free vegetables and fruit in PKU').

Write down the foods your baby has eaten, until you get used to the process of measuring and counting. Your dietitian will explain how to fill in food records, which may need to be taken to clinic visits or sent with blood samples.

Your baby's diet and blood tests will be carefully monitored by your PKU dietitian and the rest of the metabolic team. This will keep the phenylalanine levels in the safe range and ensure your baby is growing.

Meal planning: balancing counted protein and free foods

As new foods are added, the task of meal planning begins.

- Aim to spread out protein throughout the day.
- Decide how many grams you have for each meal and snack.
- Choose one counted protein food such as rice cereal, potatoes, peas or corn.
- Choose uncounted vegetables or fruits such as carrots, green beans, peaches or pears.
- Measure the amount of counted protein food your child may have to add up to the number of grams available for that meal.
- If your child doesn't eat all the food offered, estimate how much is left and subtract it from the calculated grams. You may need to account for this in the next meal or snack.
- As your child eats more foods, they will need less breast milk or standard infant formula.
- Be sure to let the dietitian know if your child is not eating all of the grams – the formula may need to be adjusted.

'We manage the diet by having some foods we can all eat and then something similar to our food that fits the PKU diet. So, if we have spaghetti bolognaise, my son has low-protein pasta with tomato sauce, and we all have the salad.'

You may talk about foods as: 'yes', 'no' or 'maybe'.

'Yes' foods are free and very low in protein, such as apple, carrots and low-protein products. 'Maybe' foods must be measured, such as crackers, cereal, potatoes, some vegetables and fruits. 'No' foods are very high in protein, such as any kind of animal or fish meat, eggs and dairy foods.

Feeding your child from one year onwards

By now, your child is probably eating fruit and vegetables, and a variety of low-protein cereals, bread, pasta and biscuits. Here are some other suggestions:

- offer small pieces of suitable soft-cooked family foods
- give most drinks by cup
- have set meal and snack times
- count the protein carefully
- offer a set number of grams of protein at each meal and snack
- continue to keep a record of grams of protein – food records may need to be sent in with blood tests and brought to clinic visits
- the phe-free formula may be the only formula your child is having at this stage.

Your child will let you know when they've eaten enough. This is the time to stop feeding, even though your child may not have finished the meal. Offer realistic serving sizes. For example, a toddler eats between a quarter and a third of an adult meal. Let them ask for more if they are still hungry.

Phe-free formula

As your baby eats more solid food and takes less breast milk or infant formula, there will be changes with the phe-free formula (called the 'supplement' after your child is 12 months of age).

The supplement replaces high-protein foods such as meat, fish, chicken, eggs, tofu, nuts and cheese. Your dietitian may adjust the quantity or type of supplement your child consumes to meet their nutritional needs as they get older (refer to chapter 4 'Understanding the PKU diet and protein supplement' for more information).

'A tip I have for changing over from one formula to another is to mix a quarter of the new formula with three-quarters of the old one, bringing it to full strength over a few days. My child's tastes are so defined, she notices the slightest change.'

Other drinks

Water is the best drink to offer apart from formulas or supplements. Keep high-sugar drinks such as juice, cordial or soft drink as a treat on special occasions. Too much of these drinks dulls a child's appetite for food, may make them overweight and isn't good for their teeth. Some plant-based milks are naturally low in phe, for example, rice, almond or coconut milk. These can be used in cooking and on cereal (check the label – protein may need to be counted in the daily allowance for some brands).

Feeding problems

Food refusal is a common but frustrating problem during the early childhood years. When children need to follow a special diet, it can make the problem even worse.

Aim to have three meals a day, with a snack mid-morning and mid-afternoon. Don't let your child eat continuously throughout the day; let your child know when it will soon be time to eat.

It is quite normal for children's appetites to vary from one day to the next. If they are growing well and phe levels are in the target range, they are eating enough. Understanding why children don't always eat as well as we would like them to makes it easier to avoid the situation where mealtimes turn into an unpleasant experience for the whole family.

It may help to allow your child to choose between two foods or be involved in preparing the food. Many toddlers want to feed themselves rather than be spoonfed. Give them their own spoon to try. Offering plenty of finger foods encourages independence.

7 tips for reducing tension at mealtime

1. Respect that your child may not be hungry

After 12 months of age children don't grow as quickly, which means their appetite won't be as large as it was in the first year. Children often compensate for reduced appetite by eating more at subsequent meals or over the following days.

2. Respect their growing independence

As toddlers start discovering that they are independent people, they may express their likes and dislikes more strongly.

3. Avoid battles over meals

Parents often become anxious when their children don't eat, and children quickly pick up on this. Some children refuse to eat, knowing it is an effective way to gain attention or their food preference.

Try to avoid offering alternative foods or extra snacks if a meal is refused. It is never a good idea to force-feed a child. This often leads to fear of mealtimes and further refusing of food.

Continue to offer new foods over time. It may take many offers before your child will taste the food, and many tastes before they like it. Praise your child for trying new foods.

Remember to stay calm if your child refuses the foods from their meal containing protein. There may be an opportunity to make up some grams later or the next day, or your child may just be experiencing a low day. The blood test results will tell you if there is a problem. Your dietitian can discuss strategies to prevent food refusal.

4. Help your child understand their special diet

A child with PKU may find it difficult to understand why they can't eat the same foods as others. As a result, they may refuse to eat their food. If possible, make your child's meals similar to the family meal. For example, if the family is having beef stir-fry and rice, serve your child stir-fried vegetables with low-protein rice. A fruit and vegetable share plate in the middle of the table is a way to get all family members involved.

Try to offer simple explanations to your child about their need for a special diet. These explanations will become more complex as they grow older. Ask your metabolic clinic team or other parents about approaches they have found helpful.

'It was a pain to get my two-year-old son to eat. I'd prepare all this special food for him and he'd just push it away. The metabolic team suggested I give him his food before his supplement when he was really hungry. I let him get down from the highchair and run around for half an hour, then I sat him on my knee and read him a story while he drank his supplement from his cup.'

5. Create a relaxed mealtime environment

Parents and siblings can be good role models for young children. Eat meals together as a family as often as possible.

Keep the mealtime atmosphere relaxed so that this is an enjoyable time for the family. Turn off the television, tablets and mobile devices and engage in positive conversation at the dinner table.

6. Keep regular routines

Children respond well to having predictable routines. They need to eat regularly to meet the demands of their growing bodies.

Children have short attention spans. Set aside 20–30 minutes for meals, and 10–15 minutes for snacks. Give a five-minute prompt before the meal to finish playing and wash hands. Seat your child at the table for meals when the food is ready. Forcing your child to sit for longer may lead to further refusing of food.

7. Keep a positive attitude to your child's diet

Allow your child to form their own opinions about their diet. Stay calm and avoid becoming stressed or frustrated.

It is important for your family and friends to have a positive attitude towards your child's diet. If others tell them their diet is awful, it makes it harder for them to take the formula or eat their special foods.

Resources

For more information on feeding your child, see the following websites:

- https://raisingchildren.net.au/
- https://www.ellynsatterinstitute.org/

8

Encouraging independence and talking about PKU

- Encouraging independence
- Guide to encouraging and supporting your child's independence at different stages
- Activities and role plays
- Transition to adulthood

Encouraging independence

When your child is very young, you have the main responsibility for managing their condition – planning meals, taking blood tests, adjusting diet according to phe levels and explaining PKU to other people. As they grow up, it is important to start sharing this responsibility with your child.

Helping your child manage their PKU has a number of advantages for them. It:

- encourages acceptance
- helps them develop a good **understanding** of PKU
- increases their **confidence** in managing their condition
- helps you and your child **work together** better as a team.

When children with PKU are able to accept and take some responsibility for managing their condition in everyday life, long-term adjustment and dietary control tend to be better. This helps them prepare for adult life when they will need to take full responsibility for their PKU.

As your child grows, your role will change from primary manager to supervisor or coach. Later, you will observe and support your child as *they* make the important decisions.

When discussing PKU, the low-protein diet and the supplement with your child and others, use positive language. This supports your child and nurtures a positive attitude to treatment from the beginning.

Guide to encouraging and supporting your child's independence at different stages

The following tables are a guide to how you can support this transition and talk to your child at different ages.

Age	What you can expect your child to do/know	What you can do to support your child
Toddlers: 2–5 years	• are aware that they have a special diet • know they need to take their supplement and watch you prepare it • know they need to have blood tests and come to clinic • check new foods with their parents/carers • start to learn yes/no foods • are aware that the protein in food is counted • willing to try new low-protein foods	• use simple examples to explain PKU and the diet. You can explain that everyone is born with different qualities, PKU is one of them. Starting this concept early builds acceptance of the condition from the beginning • a storybook to assist can be found at the following link: https://www.hgsa.org.au/resources/asiem-resources-for-parents-and-families/robin-and-i-explain-pku • teach them to ask you if they can have unfamiliar foods and that they can say 'no' when offered food by others. Learning their diet is different can set them up for success when they go to preschool and daycare • stay positive and focus on the foods they can eat • avoid showing that you dislike the taste of the supplement or low-protein foods • encourage them to assist with simple aspects of food preparation such as mixing ingredients in a bowl • attend clinic regularly so that your child knows that is part of routine PKU care

Age	What you can expect your child to do/know	What you can do to support your child
Early childhood: 5–8 years	• understand that they have a condition called PKU • understand that they can't eat high-protein foods • have a basic knowledge of why the supplement is important and help you prepare it • understand why the blood test needs to be taken and if ready, can assist in taking their own blood tests • practise using scales to weigh foods and count protein • have ideas about meal choices, help prepare meals and continue to try new foods • start learning to select low-protein foods in social situations	• talking about goals, weekend activities, sports and how they relate to PKU can make the need for appropriate phe levels more meaningful • help your child to make decisions about aspects of their diet. For example, taking supplement to school, or whether to take food to a social event or choose allowed food from what is provided • encourage them to assist with food preparation • encourage your child to talk openly and honestly about what they eat at school, if they eat something that isn't normally in their diet or if anything was said that upsets them. Keep calm and troubleshoot how to deal with this • attend clinic regularly and attend education days or events run by metabolic/PKU organisations • encourage discussing PKU with friends so they understand or can help offer support in social situation • some children like to do a class presentation on PKU, offering classmates the chance to understand parts of the PKU diet and supplement • make your child aware of other people you know who follow a special diet, so they know they aren't alone. PKU support groups, social events and camps can assist. Refer to chapter 19 'PKU resources' for more information
Late childhood: 8–12 years	• have a basic understanding of PKU, phe levels and low-protein diet • understand that the supplement helps provide nutrition and lowers phe levels and they are able to get it out or prepare it • practise taking their own blood samples and have an idea of how frequently they need to take blood samples • can read and calculate the grams of protein from product labels with assistance • can weigh foods accurately and count the grams of protein • are able to make appropriate meal choices most of the time and prepare simple meals	• all as above

Age	What you can expect your child to do/know	What you can do to support your child
Late teens: 14–18 years	• have a thorough understanding of PKU and its effect on the body • can explain how PKU is inherited and the likely inheritance in their own children • prepare/take their own supplement with little prompting • can measure foods and count protein accurately • are able to make dietary changes depending on blood phe levels with support • are able to make appropriate meal choices • can prepare appropriate meals • remember to take their own blood samples and send them in for testing • know how to order supplements and low-protein food • know how often clinic visits are needed and start to take responsibility for making their own appointments • know how to contact their metabolic clinic team when required • are aware of the pregnancy risks associated with maternal PKU • attend an adolescent metabolic clinic if available or have a plan in place for transition to an adult metabolic service • start to talk to the metabolic clinic team by themselves about their PKU management and concerns	• talking about goals, future ambitions, careers, sports and how they relate to PKU can make the need for appropriate phe levels more meaningful • provide encouragement when they take on responsibilities of PKU management • attend regular clinic appointments and encourage attendance at education day and support group events • give them time to talk about their feelings towards PKU without the pressure of discussing blood phe results or what they have eaten • constant battles over diet and supplements can cause stress for any parent/adolescent. Make sure you have time to yourself as well • if you notice deviations from diet or supplements, social withdrawal, changes in behaviour, mood or attitude to PKU discuss this with your metabolic clinic team. Support is available

Activities and role plays

You can help build your/your child's understanding of PKU using games and role plays. For example:

- placing different foods into 'yes' and 'no' or 'high', 'medium' and 'low' protein categories
- a star chart/reward system for taking blood tests, drinking the supplement and managing the diet
- putting phe results on a wall chart so they/you can monitor their/your own progress
- practise cooking low-protein recipes
- count protein together using a chart on the fridge or wall.

It's normal for you or your child to be asked questions about the low-protein diet. Rehearsing answers and scenarios beforehand may help when you are asked questions.

Frequently asked questions and example answers include:

- Why don't you eat meat?
  - I'm on a special diet.
  - I don't mind not eating meat. I'm used to not having it.
- Are you vegetarian?
  - My diet is like a vegan diet but stricter and with a supplement to ensure I am healthy.
- Are you sick?
  - No, I'm not sick. I'm healthy and my diet keeps me healthy.
- Can I catch it?
  - You can't catch PKU, you have to be born with it.
- How can you drink your supplement? It smells.
  - I have taken a supplement since I was a baby, so I'm used to it.
  - I'm used to having the supplement. It's like medicine – if you need to take it to be healthy, you take it.

Exposure to these questions and new social scenarios poses new challenges for your child. Empathise with their struggles. For example:

- 'It's really hard taking your PKU supplements every day!' Then gently remind them that taking the supplement will keep them healthier in a way relevant to them. For example, 'It helps you to be strong for football.'
- 'I don't blame you for being upset and mad that you can't eat the sausage sizzle! I would be too! As hard as this is, at least you have your own special treat to take to the BBQ.'

Transition to adulthood

Transition from paediatric to adult PKU services will be much easier if you/your child do the following:

- practise cooking low-protein recipes and assembling meals and snacks
- practise explaining PKU and communicating health care needs
- keep a record of appointments, medical history and medications prescribed
- record the metabolic clinic team names, phone numbers and email addresses on a smartphone, tablet or in a diary
- begin to make your/their own medical appointments
- write down questions for the metabolic clinic team before your visit
- have parents, friends, partner or support person remain in the waiting room while he/she spends time with the metabolic clinic team on their own
- learn new information about the health care needs of an adult with PKU
- learn the practical aspects of PKU management. For example, how to order PKU supplements and low-protein food, monitor blood phe levels, count protein in diet
- if you're more comfortable, you can consider having someone with you for support, like a friend, parent or someone else you trust.

(Transition checklist adapted from National PKU Alliance, My PKU Binder, www.npkua.org)

9

Preparing for childcare, early years education and school

- Planning
- Discussing PKU. Who do you need to inform?
- Food at childcare, preschool and school
- Taking the protein supplement
- Parties and cooking days
- What to discuss with your child

Planning

Starting childcare and preschool is an exciting time for any family. It can, however, also be stressful, and it may seem overwhelming with PKU to think about. This is completely normal.

With planning, children with PKU can easily make this transition and keep their PKU well controlled. There needs to be ongoing support and communication between the family, facility and metabolic clinic team.

Each centre and school is run differently. The following suggestions are a guide for planning your child's PKU management. Most clinics will also have a dietitian or nurse available to talk to the centre or school, to help with planning and provide information on PKU.

Discussing PKU. Who do you need to inform?

When children are young, the PKU diet needs careful supervision. You should inform the following people about your child's PKU:

- the principal, admissions staff and preschool supervisor (when you're planning your child's enrolment)
- the class teacher
- the school nurse – for support and as an education resource
- the supervisor of the childcare centre, and before or after school care
- the canteen supervisor or cook.

'When you're telling teachers your child has PKU, you're very aware of the fact that you don't want to label your child as potentially having brain damage, because if they've heard of PKU at all, they only know about brain damage. So I quickly tell them she is normal like everybody else, but that she is normal because she's eaten the right food.'

What to discuss with staff

Anyone caring for your child needs to know that:

- PKU is an inherited, non-contagious condition
- children with PKU cannot break down an amino acid called phe, found in all protein foods
- all children need a certain amount of phe for growth and repair of the body, but in PKU the extra phe builds up and can damage the brain
- staying on a protein restricted diet keeps the phe levels in a safe range which will allow the child to develop normally
- eating the wrong foods will not make them immediately sick but it will have a detrimental effect long term
- a child with PKU has a very specific diet that is calculated by a specialist team, with food portions measured out daily by the child's family. A system must be in place at childcare, preschool/kindy and during the early years at school to supervise the type and amount of food they eat during the day. As your child gets older, they will be able to monitor this themselves
- parents must be informed if the child has eaten food that is not allowed, or does not eat foods that are sent from home.

Your metabolic clinic team can provide a letter which offers a simple explanation of PKU for childcare, teachers and parents of classmates.

Food at childcare, preschool and school

Each centre or school will vary in what they provide in terms of meals or canteen food. You will need to decide whether to:

- provide all the food your child will eat while they are there
- provide the main meals but use the centre's meals or school canteen for snacks, such as fruit, salad plates or ice blocks. You could also give the centre a list of foods that are 'free' (contain minimal or no protein)
- use only the meal services provided, by either pre-ordering the low-protein foods from the menu, or providing low-protein bread/pasta/rice for the service to make into sandwiches or meals with low-protein fillings.

You are the expert in meal planning and protein counting. Often, keeping it as simple as possible for the centre or school is the best way to stay on track with your child's protein allowance.

'You can't depend on others to protect your child; you have to protect them yourself. When they're little, if they eat the wrong food and get away with it, it becomes hard for the child and the parents. At one stage I found out my child was swapping some of his lunch for another boy's cheese. When he got home we discussed it and bought some low-protein cheese to have on his sandwich. The lunch swapping stopped.'

The school needs to be reassured that, apart from needing a special or restricted diet, your child is perfectly healthy and developing normally.

Taking the protein supplement

Your child should take their supplement to childcare, preschool or school to help spread the intake throughout the day. Starting this routine early can normalise the supplement as part of school life. If your child is concerned about questions from peers, it can be beneficial for them to take the supplement in covered drink bottles, as pre-made drink packs (if available) or as a bar or pudding style supplement. Some children prefer to go to the office to have their supplement, while others prefer to have it in class with their peers.

'My son was embarrassed taking his supplement drink to school. He was much happier taking a supplement bar, unwrapped and placed in a plastic container so it looked like it was from the supermarket, saving his drinks for home.'

Parties and cooking days

Where possible, prepare in advance. Ask the teacher to notify you before food or cooking activities so you can send similar PKU-friendly foods. Examples include:

- a low-protein sausage or vegetable patty for BBQ events
- low-protein cupcakes – keep a supply pre-made in the freezer, low-protein biscuits and lollies to keep with your child's teacher.

'I arranged with the teacher to keep some low-protein cakes in the staff freezer. She takes a cake out in the morning when the birthday child brings their cake in, and it defrosts by recess when they are ready to celebrate.'

See chapter 10 'The PKU diet' for more ideas about food for special occasions, school camps and excursions.

What to discuss with your child

- Talking to your child is an important part of preparing for childcare, preschool or school. While the school staff will do their best to supervise, you will feel more secure if your child understands and is able to manage their diet themselves.

Things to talk to your child about include:

- foods which are allowed and those to avoid
- bringing home uneaten food in the lunch box so that you can calculate grams of protein
- buying only low-protein food from the canteen (and no diet drinks or other foods with artificial sweeteners that are not allowed on the diet)
- not swapping lunches with friends
- deciding when to drink the supplement
- how to explain the different diet to other people
- what to do about teasing about the diet
- if they don't eat their food or they eat something they wouldn't normally eat, that they need to talk to you about it, so that you can adjust dinner or the next day meals.

'I find I am constantly watching her to make sure she doesn't eat the wrong thing. Kids will be kids and it does happen. The important thing is not to stress about it. I find talking to her about what she can and can't have is the best. She's almost four now and she knows what she can and can't have.'

Children often find it difficult being different from others and they feel embarrassed about having to eat different food. Refer to chapter 8 'Encouraging independence and talking about PKU' for more information.

10

The PKU diet

- Family mealtimes
- What makes up a PKU meal?
- Preparing a low-protein meal and family meal together
- Desserts
- Lunch ideas
- Snacks
- Recipes
- Getting organised for camps and excursions
- Children's birthday parties
- Christmas, Easter and other special events
- Teenage and adult parties
- Barbecues
- Eating out
- Ingredients with hidden protein or phe
- Healthy choices
- Controlling your weight

Family mealtimes

Family mealtimes provide a good opportunity to talk about the day and also for children to learn about healthy food choices. From around one year of age, it's important to include foods the whole family can eat at mealtimes. This helps your child to feel part of everyday family life.

Ways of doing this include:

- base the meal on vegetables, low-protein breads, cereals, rice or pasta prepared in a similar way to the family meal. The picture below shows the different parts of a low-protein meal
- the chart below suggests ways to prepare the family meal and the low-protein meal at the same time
- encourage your child to be involved in preparing meals. As they mash the vegetables, stir the mixture and add flavourings they will be learning about the foods they can eat and enjoy.

What makes up a PKU meal?

Preparing a low-protein meal and family meal together

See the following two tables showing family meals and how to adapt them so they are low protein. Adapt the foods in the PKU meal column to provide the protein required at that meal. For example, you can lower the protein content using egg replacer, low-protein cheese or low-protein pasta, rice and flour in place of regular varieties. Then add the required amount of protein from counted foods such as peas, sweetcorn, potato or gluten-free products.

Family food	PKU meal
Roast meat with roast vegetables and gravy	Roast vegetables: •with a low-protein sauce or gravy and low-protein garlic bread or buns •rolled in a low-protein pancake •with low-protein bread and a sweet chilli dipping sauce •stirred through cooked low-protein pasta or rice •add in new flavours e.g. balsamic glaze, garlic, mixed herbs, Mexican, Cajun or Moroccan spice mix •baked sweet potato boats topped with zucchini, capsicum, eggplant, grated low-protein cheese and salsa •roast a whole cauliflower with a spice rub, e.g. paprika, garlic, olive oil, salt
Quiche or pie	Grated vegetables (zucchini, parsnip, carrot, etc.): •baked as a pie with herbs and a low-protein crust •with a low-protein pasta sauce stirred through and a low-protein bread crumb topping •made into a vegetable quiche using egg replacer •piled into a vol-au-vent shell
Grilled meat and steamed vegetables	•Vegetable kebabs: capsicum, onion, mushrooms, zucchini grilled on skewers •Vegetable patty and steamed vegetables •Fried or baked crumbed zucchini or eggplant slices and steamed vegetables
Stir-fry with rice or noodles	•Stir-fry vegetables and low-protein rice or pasta with a sauce, e.g. sweet and sour sauce •Cook the chicken or meat separately, and use the vegetable stir-fry for everyone
Sausages, mashed potato or chips and salad	• Low-protein vegetable sausages, mashed potato or low-protein bread and salad
Beef patties in hamburger bun or roll	•Vegetable patty in low-protein toasted bread or bun
Tuna mornay or macaroni cheese	•Vegetable macaroni bake: herbs, low-protein pasta, low-protein cheese and a white sauce using low-protein milk, rice milk or a stir-through pasta sauce, baked with a low-protein bread crumb or sliced tomato topping
Pizza	•Low-protein pizza base made on low-protein flour or gluten-free wrap, spread with tomato paste and topped with olives, pineapple and grilled vegetables, e.g. eggplant, zucchini, capsicum and low-protein cheese •Use low-protein bread rolls to make pizza subs
Barbecue	•Vegetable and fruit skewers barbecued with low-protein bread or pasta salad
Garlic bread	•Low-protein rolls cut and spread with garlic butter, or cracker biscuits spread with garlic butter
Spaghetti bolognaise and salad	•Low-protein spaghetti with cooked vegetables, herbs and tomato puree or a stir-through low-protein pasta sauce and salad
Mexican – beef nachos or tacos	•Vegetable 'chilli no carne' – capsicum, onion, zucchini, corn and tomatoes cooked with Mexican spices. Serve with low-protein rice or fill tortillas made with low-protein flour or use gluten-free wraps •Nachos – cut low-protein tortillas into triangles and bake to make tortilla chips •Baked cauliflower pieces coated with low-protein breadcrumbs and spices served in low-protein tortillas with salad and chipotle mayonnaise

'A quick meal in our house is a pack of stir-fry vegies from the supermarket and the sauce is a soy, honey and garlic marinade.'

'I find the gluten-free products in the supermarket are worth checking out. Some are really high in protein, but some aren't. I shop in the Asian section too as some of the noodles and biscuits are low in protein.'

Desserts

'For a quick dessert we purchase meringue nests, fill them with whipped cream from a can and put sprinkles or fruit on top. Yum!'

Desserts can be included in the low-protein diet in moderation. Some examples are:

- low-protein breads and cereals in the dessert if they were not part of the main low-protein meal
- apple (or other fruit) crumble – make topping from low-protein crumbed sweet biscuits or low-protein flour, sugar and margarine and baking powder
- low-protein pancakes or crepes with fruit, honey, lemon juice or cinnamon
- low-protein rice or pasta with sugar or honey and margarine, cream, rice milk or low-protein milk
- low-protein rice with fruit and coconut cream or regular cream
- sago or tapioca with various flavours
- custard made from low-protein milks and custard powder served with fruit
- churros made with low-protein flour
- sorbet
- whole baked apple – topped with butter, brown sugar and cinnamon
- coconut yoghurt parfait – layered with diced fruit, sprinkle with crushed low-protein biscuit.

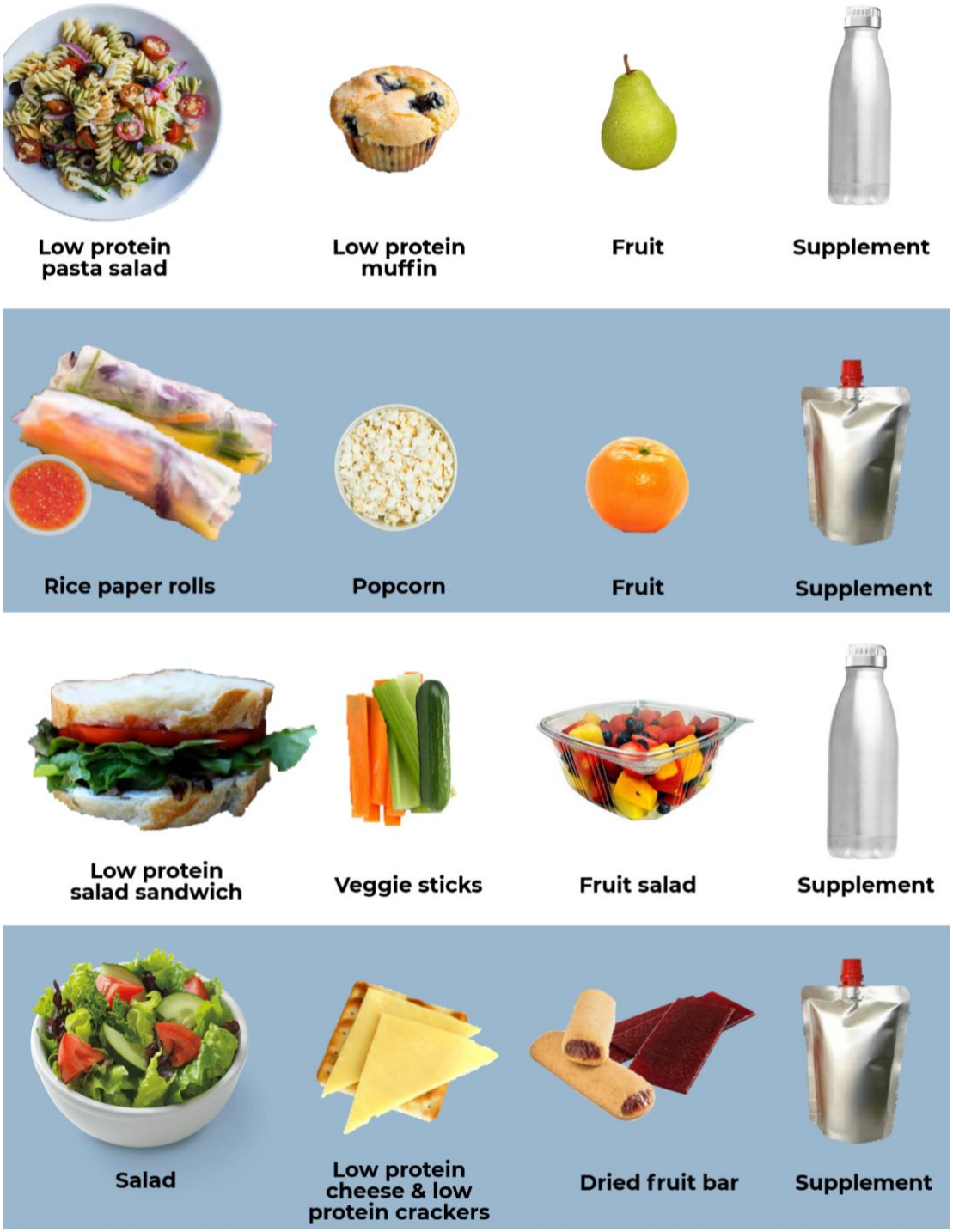
Lunch ideas

When you're preparing lunch for your child or yourself, remember that variety is enjoyable. Providing an appetising packed lunch is a good way to help prevent the temptation to swap lunch with someone else or purchase lunch out. See the following tables for lunch ideas and lunchbox combinations:

'For me, taking food from home is the easiest way to keep track of my protein intake during the day. If you are buying lunch, do your homework on shops near your workplace, college or university and build a relationship with them so they will know your favourite foods and be able to adjust the food to your dietary requirements.'

Low-protein sandwich fillings	Salads and soups	Bread and fruit
• avocado • grated carrot with sultanas • shredded lettuce • tomato and cucumber • beetroot • gherkin relish or chutney • olive tapenade (paste) • leftover chargrilled vegetables • cold vege burger/patty/ sausage • banana and cinnamon • low-protein cheese and salad/chutney • jam/honey	• cherry tomatoes • cucumber sticks • carrot sticks • stuffed olives • sprouts • celery sticks • lightly cooked cauliflower sprigs • low-protein pasta or rice • salad, e.g. homemade basil pesto without nuts or cheese • leftover grilled/baked vegetables with lettuce leaves and dressing • pumpkin, vege minestrone or tomato soup	• fresh fruit (cut up and put into sealed containers) • individual fruit snack packs, fruit bars or dried fruit • low-protein bread rolls • low-protein crispbreads • rice cakes • cold pizza slice • low-protein muffins, pikelets, sweet biscuits or cake

Example lunchbox combinations



Snacks

Low-protein snack ideas include:

Fruit and vegetable based	Supermarket products	Home made using low-protein ingredients
• avocado • grated carrot with sultanas • shredded lettuce • tomato and cucumber • beetroot • gherkin relish or chutney • olive tapenade (paste) • leftover chargrilled vegetables • cold vege burger/patty/ sausage • banana and cinnamon • low-protein cheese and salad/chutney • jam/honey	• popcorn • vege chips • fruit ice blocks and icy poles • rice, almond or oat milk shakes or smoothies • savoury crackers • gluten-free breakfast cereal with rice milk • coconut yoghurt • low-protein cheese, olives, gherkins and crackers • gluten-free pretzels • puffed rice bars • dried fruit straps and bars • gluten-free wafer biscuits •	• low-protein baked goods, such as scones, biscuits, cupcakes or pikelets • sandwiches • low protein pasta made into a soup • low protein breakfast cereal with rice milk • mini pizza sub on low protein bread with olives, capsicum and low protein cheese

Recipes

Cooking for a low-protein diet takes time to learn. Early results may not live up to your expectations, but it gets easier. Most people find that before long they're compiling their own list of favourite low-protein recipes and inventing new ones as they gain experience.

Recipes and protein content

You need to count the protein in the recipe as you would if you ate the foods individually. For example, if you regard most vegetables as 'free', and only count those such as potato, sweet potato, corn and peas, then do the same when they're in a recipe. Adapt the recipes according to how strict a low-protein diet you are following.

How to adapt a recipe

Here are some tips on adapting recipes for the low-protein diet:

- recipes based on vegetables, salads, vegetarian, vegan, fruit, potato, rice, pasta and noodles are more likely to be suitable
- leave out the meat in meat and vegetable dishes
- use a low-protein milk substitute for milk, e.g. rice, almond or oat milk
- where a recipe includes a white sauce, use your own low-protein sauce
- use egg replacer or other egg substitutes, e.g. apple puree in muffins, instead of egg
- use low-protein cheese or stir in a spoonful of coffee whitener in recipes using cheese such as risotto
- use maize cornflour or low-protein flour instead of plain flour
- use low-protein or gluten-free pasta or rice instead of regular varieties
- canned or fresh jackfruit can be cooked to make low-protein vegetarian meat substitutes, e.g. cooked in BBQ sauce to make 'pulled pork'.

'I didn't want PKU to stop my child doing anything. My daughter was seven when she first went on a four-day school camp. I was weeks getting everything ready and planning it. I sent a book along with her so her teachers would understand how her diet works. She'd add up the protein and I'd check it when she got home.'

Where to find recipes

Many companies that sell low-protein foods as well as PKU community groups have recipe and food ideas on their websites. Many personal blogs, Facebook groups, Pinterest boards, Instagram accounts and YouTube channels are great places to find low-protein food ideas and recipes. Be sure to carefully check the protein estimation in the recipe.

Tip: You may find more low-protein recipes by including 'PKU' in your search terms. Suggested Instagram tag search terms: 'lowproteindiet', 'lowproteinfood', 'pkufood', 'pkudiet'. Refer to chapter 19 'PKU resources' for information on low-protein food company and recipe websites.

Getting organised for camps and excursions

A low-protein diet can be easily managed while your child is on camp or excursions. For a day trip, pack the usual lunch and supplement, but for longer excursions a bit more planning is required.

- Discuss your requirements with the camp supervisor, including the need for a supplement and give them a clear meal plan.
- Obtain copies of the menus in advance so you can help plan your meals and adapt them as necessary.
- Take ready-made supplements such as those in cartons, puddings or bars. If using a powdered supplement, pre-pack the prescribed number of scoops into separate containers so that it just needs water added. Pack extra in case of delays.
- Take a supply of low-protein bread, pasta, biscuits and low-protein milk.
- Include some free foods for unplanned snacks or if hungry.

If you're going camping with friends, plan in advance what you will take. If someone else is organising the food, tell them what you can and can't eat. Take plenty of low-protein food. Some ideas are:

- dried fruit, fresh fruit/vegetables or individual fruit snack packs
- low-protein noodles and pasta and ready-made pasta sauces
- low-protein cereal and milk e.g., rice, almond, oat or specialised protein-free milks
- snack bars, low-protein biscuits and crackers, rice cakes or corn thins
- vege chips – packed in a plastic container so they don't get crushed
- lollies (check the label for lower protein options) or low-protein chocolate (available in Australia).

Children's birthday parties

Before the party

Parents of children with PKU worry that their child will eat too much protein at a party. Here are some tips for managing your concerns:

- to help minimise the risk of overeating, make sure your child doesn't miss any meals or snacks before the party
- give fewer grams of protein at the meal before the party to leave a little room for party extras
- avoid major discussions about food in front of your child and their friends
- be realistic, and expect that your child may have more protein than usual. If needed, you can lower the protein content of the meals or snacks following the party to help compensate for this
- take a plate of low-protein food to share – let your child help decide what foods will be included on the special platter. Try to drop the food off before the party so your child doesn't feel self-conscious about eating different food
- explain to the host of the party that your child is on a special diet when you accept the invitation, rather than waiting until the day of the party. Discuss suitable foods and ask if they want some specific suggestions
- deliver a PKU cupcake to the host before the party so your child can have this when others are having birthday cake
- tell the host that your child should not have diet soft drinks, diet cordials, milk shakes or flavoured milk – water, regular cordials and soft drinks are suitable alternatives.

Parties at home

'When you have the party at home, it's easy to cater for low-protein party food. The fairy bread was made from low-protein bread, the "sausage rolls" were low-protein pastry filled with vegies, and we had some vege chips.'

Plan the food well in advance, involving your child in choosing the menu. Include some low-protein options at the party for everyone to share.

Here are some tips to help make the day a success:

- tell parents of your guests that your child is on a special diet and let them know that presents containing food would not be appropriate for our child
- include lots of regular foods that are low in protein for everyone – include some of your child's favourite low-protein foods as well as some regular party favourites for the guests
- use websites, magazines or recipe books for ideas – many party recipes can be adapted so they are suitable for PKU
- tell your child before the party about which foods they can eat – try putting fun markers, such as little paper flags and cocktail umbrellas, on the foods they can eat
- games and activities can take the focus off food. If the game involves food try substituting low-protein foods for the regular food, for example low-protein chocolate, and low-protein lollies for a treasure hunt
- use non-food prizes for games, e.g., stickers, pencils, hair accessories, bookmarks, diaries. Low-protein lollies, small packets of chips/vege chips or biscuits can be used for the lolly bags.

Making a PKU birthday cake

'Sometimes I'd form the birthday cake with little cakes and his had special icing so he knew which ones he could eat so it looked like he was having the same as everyone else. There's a lot of psychology involved.'

Party books will give you ideas for shapes and decorating. Here are some ideas for the cake:

- use a favourite low-protein cake recipe and decorate it with icing and low-protein lollies
- make a low-protein ice-cream cake or find a ready-made one with low-protein content. Some gelaterias make cakes from sorbet
- make a jelly mould using low-protein jelly, set fruit into the mould or pile it on top and decorate with whipped cream
- bake a two-tiered cake, with a regular cake on the bottom and a smaller low-protein cake on top, and cover all with icing and decorate – serve the guests from the bottom and your child from the top
- make shape cakes, for example houses or animals, and use the PKU cake for easily identifiable parts such as the roof
- cut watermelon into the shape of a cake, top with icing or coconut yoghurt
- make 'honey joys' with cornflakes, honey and butter, or 'chocolate crackles' in patty pans and display on a cupcake stand
- make low-protein donuts and pile on top of each other to make a pyramid.

'My daughter would come home after birthday parties with her piece of birthday cake and lolly bag and auction them off to her big sister and brother, who would buy them with jellybeans (which she could eat) and coloured pencils.'

'By the time he was five he refused to let me take special birthday foods to friends' parties. He'd just go and have fun.'

Trusting your child

Parents of very young children often ask the host whether they can stay at the party and help out. This helps them to keep an eye on their child.

By school age, most children are attending parties on their own and there is no reason why this can't be the case with a child who has PKU. Most children of this age can identify high and low-protein foods and it is a matter of parents trusting them. If they've shown they can do this at home, they're likely to carry it through when they are out socially.

Christmas, Easter and other special events

Find special alternatives to the usual foods that are part of your family's or your friends' celebrations.

For Christmas, bake some low-protein fruit mince pies and low-protein Christmas pudding or cake. You can make low-protein Easter eggs by filling plastic casings from craft stores with melted low-protein chocolate. Try melting low-protein chocolate and a small quantity of dark chocolate together and putting them into different moulds. Shop around for some of the cheaper chocolate eggs that are lower in protein. Try filling Easter bags with protein-free lollies.

'I buy "natural" carob and milk-free Easter eggs which are low in protein and sold in major supermarkets. These little eggs can be melted down and made into bigger eggs or shapes. I buy packets of them at the end of Easter and use them for treats in the following months, as they have a fairly long use-by date.'

'We planned her Christmas menu in advance: vege chips, fruit and low-protein lollies for nibbles, and the vegies from the baked dinner weighed of course and grams calculated, with a vegie rissole. We all had instant gravy rather than pan juice gravy, as the packet gravy is lower in protein. Her grandmother made her a low-protein pudding and I made custard using custard powder and Coffee Mate.'

Teenage and adult parties

Eating low-protein foods in the time leading up to the party should give you enough protein to enjoy the party without going overboard. There will be times when you go over your intake of protein. If that happens, make sure you eat less protein for the next day or two.

If you're going to a party, taking a dish along helps to avoid questions about your diet. Advising your hosts of your food restrictions is a great way to plan for parties. Firstly, food can be made for you, and secondly, finding out what's on the menu means you can create a low-protein dish similar to the food the other guests are having.

Remember: it's ok to say no when someone offers you food.

Barbecues

Here are some ideas to include at barbecues:

- homemade vegetable and fruit skewers
- vegetable kebabs marinated in garlic and honey sauce or other low-protein marinades
- hash browns
- commercial vegetable fingers
- homemade vegetable patties
- homemade vegetable sausages
- corn on the cob in foil
- mushrooms
- eggplant slices
- roast vegie salads, green salad, low-protein rice/pasta salad
- dips, e.g. guacamole, salsa, eggplant with low-protein crackers or bread.

Eating out

General tips

- Look at the menu in advance to ensure there is something you like that can be adapted for a low-protein diet.
- Explain your diet to the food outlet, chef or owner. Many places are happy to cater for different dietary restrictions and adapt menu items accordingly.
- If you eat regularly at a venue, take low-protein food such as pasta/rice – the venue may be happy to cook it for you with their own sauce.
- Create your own meal – dining options with 'build your own' salads or side dishes can be more flexible.
- Many takeaway food outlets label their food with the nutrient content so you can work out the protein content.
- Be mindful of how much you are consuming. Meal sizes are often larger when eating out, making it easy to eat too much protein.
- Don't be afraid to ask questions about the ingredients used in the meals.

Ingredients with hidden protein or phe

Ingredients	May Contain
Caesar dressing	Anchovies
Gravy or soup broth	Meat or poultry
Miso	Soybeans
Satay sauce	Peanuts
Soy sauce	Soybeans
Tahini	Sesame seeds

Cooking for family and friends

Adapting low-protein meals for those who do not have PKU can be straightforward. You will need to add a source of protein, such as red meat, chicken, fish, eggs or cheese. Vegetarian meals are suitable, but you will also need to include protein, such as cheese, tofu, nuts or legumes (lentils, chickpeas, red or white beans). Use regular bread and pasta. Low-protein PKU meals are not suitable as a main meal for someone on a non-PKU diet because they do not have the PKU supplement as a source of protein to go with the food.

Healthy choices

Fat facts

Some fats are good for the body, others should be eaten in moderation. The balance of the types of fats eaten can impact heart health. They change the balance of good and bad cholesterol in the blood. If there is heart disease in your family, or you have high cholesterol levels, talk to your dietitian about which fats you should include in your diet.

Intake of some of the healthy fats can be inadequate in a low-protein diet. Food labels give information about the amount and type of fat the food contains. See the table on the following page for more information.

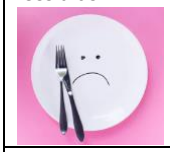
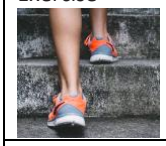
Types of fat

Type of fat	Impact on health	Food sources
Saturated and trans fats	▪ Negative impact, linked to increased bad blood cholesterol which is a risk factor for heart disease ▪ Trans fats also lower your good cholesterol level	▪ Hot chips, commercial biscuits, snack foods, cakes and pastries, cooking margarine, butter or cream ▪ Other sources of saturated fat, from animal products such as meat, are not commonly eaten in a PKU diet
Monounsaturated fats	▪ Positive impact, increase good blood cholesterol levels	▪ Extra virgin olive oil, canola oil, peanut oil, avocados, olives and some margarines
Omega-6 polyunsaturated fats	▪ Positive impact. Considered essential; the body can't make them. Positive effect on heart health and thought to have many other health benefits	▪ Oat bran, rice bran, sunflower oil, safflower oil, and polyunsaturated margarine
Omega-3 polyunsaturated fats	▪ Positive impact. Considered essential; the body can't make them. Positive effect on heart health and thought to have many other health benefits	▪ Canola, linseed, mustard seed or walnut oils, and dark green leafy vegetables ▪ Sources such as fish, seafood, eggs and nuts are usually excluded from a PKU diet ▪ Many PKU supplements provide omega-3 fats, or a fish oil supplement can be taken. Discuss with your dietitian

Controlling your weight

Tips for weight reduction on a low-protein diet

If you are concerned about your weight or growth, discuss it with your dietitian. If you need to reduce your weight, you need to consume less energy each day than your body uses.

Avoid rapid weight loss, fad diets and commercial weight loss aids 	■ In most cases weight loss needs to be gradual – no more than half a kilogram a week is more likely to be sustainable ■ Rapid weight loss breaks down muscle as well as body fat. Since muscle is mainly protein, this could increase your blood phe level ■ Fad diets usually lack nutrients and can be high in protein which is not suitable for people with PKU
Exercise 	■ Be physically active ■ Try 60 minutes of moderate to high physical activity per day ■ Try brisk walking, swimming, cycling or dancing ■ Exercise that builds muscle helps to prevent increases in blood phe levels while you lose weight
Limit screen time 	■ Keep time spent using the computer, TV, video games, smart phones to a minimum. For example, less than 1 hour per day for 2-5year old children; less than 2 hours per day for anyone over 5 years old ■ When working or studying remember to get up and move as often as you can
Supplement 	■ Take the amount of supplement recommended by your dietitian ■ If you don't take enough supplement, you're more likely to be hungry, eating more food or breaking down muscle, causing blood phe levels to rise ■ Ask your dietitian's advice on which supplement is best when losing weight as some are lower in energy
Eat regularly, including your protein allowance 	■ Eating your protein allowance will help keep phe levels in the target range. Choose foods for your protein allowance that are filling, like fruit and vegetables, rather than processed snack items ■ Include free vegetables every day ■ Carbohydrates such as low-protein or regular bread, pasta or rice and starchy vegetables such as potato and corn, should take up no more than 1/3 of your plate ■ Fruit is a great snack. Include 2 pieces per day
Limit intake of foods containing saturated fat, added salt, added sugars and alcohol 	■ Refer to the above table 'Types of fat' for more information ■ Drink water, unsweetened mineral water, soda water, tea or coffee rather than juices, soft drinks and cordial. Artificially sweetened soft drinks and cordials are another low-calorie option however ensure they are phe free (See 'Artificial sweeteners' in chapter 6)

Tips to gain weight on the PKU diet

If you are concerned about your weight or growth, discuss it with your dietitian. If you need to gain weight, you need to consume more energy than your body uses each day, by eating more food, or adding extra fat and sugar to your food.

Supplement 	▪ You may need a little more supplement than you are currently consuming. Take it throughout the day in three or more doses. Your dietitian will guide you
Have your protein allowance and eat three meals and 2–3 snacks a day 	▪ Eating your protein allowance will help keep phe levels in the target range ▪ Eat at least 3 meals and 2–3 snacks per day. Small frequent amounts of high-energy foods add up over the day ▪ Fill up on low-protein foods such as low-protein pasta and rice, low-protein bread, low-protein crackers with cheese and low-protein cereals
High-energy food 	▪ Add extra sugar and unsaturated fat to your food, such as honey/sugar on low-protein cereal, extra olive oil on low-protein pasta or thickly spread margarine on low-protein bread ▪ Refer to the previous table ‘Types of fat’ for more information
Energy supplements and drinks 	▪ If you only have a small appetite, specialised high-energy supplements may be useful ▪ Discuss options suitable for you with your dietitian
Make food in bulk and freeze leftovers 	▪ Have high-energy food available at all times. It is handy when you don’t feel like preparing food

11

PKU: Teenagers and adults

- Staying on diet
- Your PKU diet
- The PKU diet for teenagers and adults
- Exercise and sport
- Body image
- Tips for parents
- Living away from home
- Alcohol
- Safe off diet
- Starting or recommencing the diet in adulthood
- Support groups
- Pregnancy and sexual activity

Staying on diet

In Australia and New Zealand, all metabolic clinics recommend staying on PKU treatment for life to maintain good health.

Studies show that staying on diet throughout your entire life is beneficial and recommended.

Some people with PKU who have gone off their diet and then start it again say that when they are back on the diet, they:

- feel better
- look better
- think more clearly
- are less moody and find it easier to get along with others
- feel less tired and have more energy
- can concentrate
- can think clearly to study and do exams
- can work things out better, e.g. when trying to think strategically such as in team sports or playing games or at work
- find it easier to communicate with their partner/family members.

Brain scans using magnetic resonance imaging (MRI) show detrimental changes in the brain when phe levels in the blood are high. This improves when phe levels are lowered. The effect of these changes long term is not known.

High phe levels may mean, among other things, that you are not able to make judgements as well as you should – such as when you're driving a car, operating machinery or organising your life. Some people with PKU who are *not on diet* or who *stop taking the supplement* develop problems such as:

- tremors (the shakes)
- nervous system problems, e.g. behaving inappropriately, mood swings or confusion
- difficulty concentrating
- stiff or weak legs
- headaches
- nutritional deficiencies which can cause severe problems, e.g. lack of vitamin B12, iron, calcium and vitamin D.

If you stop your diet or supplement, then it is essential to talk to your doctor and dietitian so that nutrient deficiencies can be minimised, and you are fully informed of the potential negative consequences.

Your PKU diet

The diet for teenagers and adults may be more flexible (see chapter 10 'The PKU diet').

The acceptable blood phe level may be higher in consultation with the metabolic team, except for pregnant women (see chapter 13 'PKU and pregnancy'). Discuss this with your metabolic team. Teenagers and adults may be able to tolerate more phe in their diet and eat more regular (i.e. not low-protein) foods.

There is a range of convenient ready-to-use supplements which are useful for teenagers and adults.

Many resources are available to help make staying on diet easier. For information about reading food labels, see chapter 6 'Phenylalanine in the PKU diet'. For a list of recipe websites and information about low-protein products, see chapter 10 'The PKU diet'.

The PKU diet for teenagers and adults

Guidelines for eating a PKU diet (see chapter 6 'Phenylalanine in the PKU Diet' for more information)
Take the PKU supplement. This is the most important thing. Have it three to four times a day with meals so it's spread out (ideally over at least 12 hours). Count protein and know how much your daily protein goal is. Eat plenty of fruit and vegetables, as well as rice, pasta, bread and crackers – low-protein or not, depending on your protein target. Make sure you're getting enough vitamins and minerals. Take vitamin, mineral and tyrosine supplements if recommended.
Tips for healthy eating on a PKU diet
Drink plenty of water. Eat the right types of fats and limit your overall fat intake (see 'Fat facts' in chapter 10). Choose foods low in salt. Limit foods high in added sugar. Prevent excessive weight gain by being active and eating a nutritious diet (see 'Controlling your weight' in chapter 10). If you choose to drink alcohol, drink it in moderation and avoid phe-containing artificial sweeteners as mixers.

Eating a healthy diet and doing regular exercise decreases later in life the risk of health problems, such as heart disease, diabetes and some cancers. For information on how to lose or gain weight, see 'Controlling your weight' in chapter 10.

Exercise and sport

Having PKU does not limit your ability to participate in exercise or sport.

Considerations for exercise:

Fluid and exercise

- During and after exercise your body needs fluid. Drink plenty of water.
- Sports drinks are not usually needed unless you are doing moderate to high intensity exercise for longer than 90 minutes, are exercising in hot conditions, sweating excessively or participating in carnival/gala-type days.

Food and exercise

- For short (e.g. 30–45 minutes) or low intensity exercise, extra food is not usually required. Your usual meal/snacks eaten before and after exercise are enough.
- For longer or higher intensity exercise, you can optimise recovery by eating carbohydrate and protein within 90 minutes of finishing your exercise. This can be achieved with low-protein breads, rice, pasta and your usual PKU supplement.

Protein and exercise

- PKU supplement: Taking your PKU supplement as prescribed should provide your body with enough protein.
- Food protein: Regular blood phe samples can help ensure you are getting the right amount of protein from food.

Protein powders and other supplements

Commercially available protein powders that some people use to aid in muscle gain are not appropriate for people with PKU as they contain high amounts of phe.

Your supplement provides the protein your body needs and helps keep blood phe under control. Think of it as your 'protein shake'. To increase your muscle bulk, ask a qualified trainer to design a resistance training program for you. Spreading your supplement over three to four doses per day can help muscle repair and growth when training.

There are now products on the market that claim to increase energy, help you work out and burn fat. The ingredients in these supplements are not regulated like the PKU supplements and can be harmful. They should be avoided. Discuss your supplements and exercise training with your dietitian for specific advice.

Body image

How we see ourselves is called our 'body image'. Having a positive body image means feeling confident and comfortable with our body and the changes it is going through. It is about feeling satisfied with our attempts to keep healthy and strong by looking after ourselves. Looking after our phe levels and accepting PKU as a part of who we are is part of that, but there will be other pressures on us to fit in.

Having a positive body image is not about trying to have a perfect body, but about accepting the way we are and being realistic about what we can change about ourselves.

Many people worry about their weight, even if their weight is in the healthy range. Pressure to be slim, taller or more muscular comes from many sources including friends, family, the media, the internet/social media and ourselves.

Dieting to lose weight or build muscle is common, but not everyone sees their body correctly when comparing themselves to friends or to stylised, unrealistic body shapes in images and models in the media. Body image can change throughout your lifetime. When you have a positive body image it is easier to lead a balanced lifestyle and have a healthier relationship with food and exercise.

We have a choice about how we respond to pressures to have the perfect body. We can try to fit the 'ideal' portrayed in the media and spend a lot of time dieting and exercising (with no guarantee of getting the results we want). We can feel inadequate and have a negative body image. OR we can challenge the body image ideal. Bodies come in all shapes and sizes. As long as we look after ours, then that is the right shape and size for us.

How to create a positive body image
Focusing on your positive qualities, skills and talents can help you accept and appreciate your whole self. Say positive things to yourself every day. Avoid negative or berating self-talk. Focus on appreciating and respecting what your body can do, rather than how it appears. Setting positive, health-focused goals rather than weight-loss-related ones is more beneficial for your overall wellbeing. Avoid comparing yourself to others, accept yourself as a whole and remember that everyone is unique and differences are what make us special. Remember, many media images are edited for publishing purposes, making them unrealistic.

Source: https://www.nedc.com.au/eating-disorders/eating-disorders-explained/body-image/

Tips for parents

Developing a healthy body image: how you can help your teen

1. Talk about body image

Many young people feel confused or concerned about the physical changes that come with puberty. You can help by listening to how they are feeling about their body and its changes. This builds openness and shows your teen that you're taking notice of what they're saying.

It's important to let everyone in your family know that teasing about appearance is not acceptable. Teasing can have a negative influence on body image and can also lead to bullying peers at school.

2. Cyberbullying

As a parent, it's important to be aware of your teen's online access and social media use. You could talk to their school if concerned about bullying.

3. Become a positive body role model

If you show that you feel positive about your own body, it'll be easier for your teen to be positive about their body.

4. Making healthy changes

If your teen wants to make lifestyle changes, make sure it's for healthy reasons. Let them know that healthy eating and physical activity aren't just for weight loss – they're vital for physical health, now and in the future. If they are overweight or obese, approaching this issue can be difficult. However, making negative comments about weight is unlikely to help with eating and activity patterns and can result in poor body image and low self-esteem.

This information was adapted from: http://raisingchildren.net.au/articles/body_image.html Feb 2017

5. Where to go for help

- Speak to a GP or your metabolic clinic team if you're concerned about the way you or your child feels about their body.
- Children and adolescents: Contact an anonymous service like Kids Helpline – phone 1800 551 800.
- All ages: Butterfly Foundation Support Line – phone 1800 ED HOPE (1800 33 4673) or email support@thebutterflyfoundation.org.au
- Youthline (New Zealand) https://www.youthline.co.nz/body-image.html

Living away from home

Finishing school or moving away from home will be exciting and filled with new experiences. Keeping your phe levels in the target range is challenging during this new phase without your family watching over you. Contact your metabolic clinic team if you need assistance.

If you are moving to a new address or you have a new phone number, always inform your metabolic clinic team.

Alcohol

While everyone needs to know about responsible drinking, having PKU does not stop you from drinking alcohol. Some alcoholic drinks are particularly hazardous for people with PKU. Keep in mind the following when you consume mixed or pre-mixed drinks:

- spirits may be mixed with diet soft drinks which generally contain the artificial sweeteners aspartame (951) or acesulphame-aspartame (962), which contain phe
- other pre-mixed drinks may contain artificial sweeteners and are required by law to be labelled if they contain phe
- milk, egg or cream-based drinks are generally high in protein – they are not labelled in the way foods are, so it is impossible to tell how much protein they contain.

Protein in alcoholic drinks needs to be counted in the usual way.

The legal drinking age in Australia and New Zealand is 18. Consuming alcohol brings additional responsibility. Impaired judgement can lead to poor food and beverage choices and high phe levels. If you choose to drink, moderation is key for health and safety.

Amount of protein in one standard drink		
DRINK	**VOLUME**	**PROTEIN**
1 small glass wine	100 ml	0.2 g
1 nip spirits	30 ml	0.0 g
1 glass regular beer	285 ml	0.9 g
1 can light beer	375 ml	~1 g
3/4 of 330 ml bottle of premix drinks	250 ml	depends on ingredients

Safe off diet

Research shows that controlling brain phe levels by staying on diet improves the lives of many adults with PKU, but this is not always possible for some people.

There is still much we don't know about the long-term effects of being off diet, but we do know that following a low-protein diet similar to the childhood diet *but without supplements* can be dangerous to your health.

You should still attend your metabolic clinic annually for nutrition monitoring.

People with PKU who are off diet are at risk of vitamin B12, iron, calcium and vitamin D deficiencies and the negative effects that has on the body. It is common for people with PKU to find it hard to eat enough of the food sources of these vitamins and minerals (red meat, chicken, fish, dairy products and legumes), after years of not eating high-protein foods.

Talk to your metabolic clinic team about the types of foods you should eat, and how much, and find out whether you need to take vitamin and mineral supplements.

Choosing to go off diet need not necessarily be permanent. You can always choose to return to diet at some time in the future, although some people with PKU who have gone off diet say they have found going back on diet very difficult.

Remember!

- Continuing PKU treatment for life is recommended.
- Don't go off diet or stop supplement without talking to your team.
- Attend clinic once a year.
- Eat a balanced diet including protein foods and check with the dietitian that you are doing so OR
- Take supplement or vitamins and minerals.

Starting or recommencing the diet in adulthood

When returning to diet or commencing diet in adulthood there may be improvements in behaviour and concentration, and other problems such as depression and anxiety may be reduced.

Attending clinic regularly, learning about the diet, supplements and monitoring your phe levels takes commitment. It is important to have support on the journey to a healthier, more productive lifestyle.

Support groups

Support groups and networks can make a big difference in the lives of some people with PKU and others living with lifelong conditions.

See chapter 19 'PKU resources' for details of local support groups, and a number of internet sites that may be helpful.

Pregnancy and sexual activity

Babies born to a mother who has consistently high phe levels during pregnancy are at high risk of having reduced mental ability, heart defects, low birth weight and a small head. To prevent these problems, women with PKU must plan their pregnancies and aim for low phe levels before they conceive. See chapter 13 'PKU and pregnancy'.

If you are sexually active, contraception is recommended to prevent unplanned pregnancy and sexually transmitted infections. Talk to your GP for advice on the right method of contraception for you.

12

Overcoming challenges

- Deviations from PKU management
- Setting up for success from the start
- Focus on the positives
- Managing conflict around PKU management – how can I help as a parent, partner, friend or family?
- Setting goals
- Withdrawal from metabolic clinic
- Where to find more information

Deviations from PKU management

Understanding PKU management and adhering to it can be two very different things. It's difficult to follow recommendations for your PKU all the time, especially when you may not feel immediate consequences. There are different challenges at every stage in life. In reality, many people will deviate from their management at some point in their lives. Children and adolescents want to belong and are often wary of being different from their peers. Adults have the pressures of work, family and home life as competing priorities for their child's care or their own. At times this can get in the way of a PKU diet and taking supplements.

It is recommended that treatment for PKU be maintained for life. It's important to address these issues so phe levels can be within the agreed target range most of the time. Below are some common challenges and tools to overcome them.

Setting up for success from the start

A positive attitude to PKU, the low-protein diet and supplements from the beginning will positively affect your child's attitude and can stay with them throughout their lives.

Tips

- Talk about PKU and its treatment with your friends and family in a positive way.
- Explaining the condition to others can reduce your/your child's feelings of isolation or difference. Keep explanations basic and build on them as you become more confident or as your child gets older. You can role play explanations about diet, supplement or PKU with family or friends. For examples, refer to chapter 8 'Encouraging independence and talking about PKU'.
- Set the expectation that regular blood testing and clinic visits are part of life.
- Engage with the metabolic team, education days or PKU support groups. Refer to chapter 19 'PKU resources' for support group information.

Focus on the positives

It's easy to feel negative about the PKU diet and blood phe levels and your capabilities as a parent or adult, especially at times when the levels are not as good as you would like.

Tips to stay positive

- PKU is a part of your or your child's life. Focus on overall wellbeing by recognising strengths, hobbies and other positive achievements. This can build confidence and resilience towards the challenges associated with PKU.
- Emotional and practical support from carers, family, friends or your metabolic clinic team can have a positive impact on living with PKU.
- Regular blood testing and clinic visits can be encouraging when things are going well, and provide an opportunity to discuss challenges and a plan going forward when things could be improved.
- Changes in mood, behaviour, concentration and academic performance may not be related to PKU. Discuss with your metabolic clinic team if you are concerned.

Remember: PKU is a part of your life – your life is not PKU.

Managing conflict around PKU management – how can I help as a parent, partner, friend or family?

It's understandable to be upset or frustrated when your child, teenager, partner or family member deviates from their PKU treatment. If you are concerned, try the following:

- Provide a safe space to talk as a 'team' without interruptions and keep emotion out of the conversation. Anger and confrontation don't solve the problem. Remember to take a step back and go to a quiet space if you feel this way.
- Gently let them know you have noticed changes in their diet or behaviour and explain why you are concerned.
- Listen to and acknowledge their point of view and experiences.
- Avoid giving advice too quickly.
- Let them know that there is help available.
- Encourage them to talk to their metabolic clinic team and friends or family members that can provide support.

Try the following conversation starters

- Living with PKU isn't always easy. I am here for you.
- What can we do together to make it easier?
- How do you feel about your PKU management?
- Which part of your diet is worrying you? What can I do to help?

Setting goals

Getting back on track doesn't usually happen overnight. Small changes can make a big difference over time.

Tips

- Write down the goal and why you want to achieve it to boost motivation.
- Be specific. Spell out what you want to do.
- Set realistic, measurable goals. For example, if your phe level is 1,000, a short-term phe goal of <700 may be achievable. Achieving goals boosts confidence.
- Make the goal relevant to you and what you want in life.
- Include a time frame so you are motivated to achieve your goal.

Withdrawal from metabolic clinic

(Adapted from https://www.blackdoginstitute.org.au/docs/default-source/factsheets, 2018)

If you're worried that a family member or close friend is not coping with their PKU management, try talking to them supportively. Suggest they consult their metabolic clinic team. If they're reluctant to seek help, perhaps explain why you're concerned and provide specific examples of their behaviours that are troubling you.

You could offer to assist them in engaging with their metabolic clinic by:

- finding someone from clinic that they feel comfortable talking to
- discussing aspects of PKU that they are finding challenging and reminding them that their metabolic clinic team can assist. For example, if taking supplements is a challenge, the PKU dietitian may offer other options or strategies to improve flavour
- making an appointment for them on their behalf
- taking them to the appointment on the day
- accompanying them during the assessment with the metabolic clinic team if appropriate
- discussing the transition from paediatric to adult services early.

Where to find more information

Australian websites

https://www.biteback.org.au/MentalFitnessChallenge

BITE BACK is an online positive psychology program with activities aimed at improving the overall wellbeing and resilience of people ages 13 to 16years.

https://www.blackdoginstitute.org.au/

Provides evidence-based resources for assistance with mental health, resilience and wellbeing.

https://www.beyondblue.org.au

Beyond Blue creates mentally healthy environments and supports people across Australia – whatever their age and wherever they live.

New Zealand websites

www.thelowdown.co.nz

The Lowdown is a website to help young New Zealanders recognise and understand depression or anxiety. It also provides information on a free 24-hour helpline.

www.justathought.co.nz

Just a Thought offers evidence-based Cognitive Behavioural Therapy (CBT) online and is designed for people with mild to moderate symptoms of anxiety and depression. CBT teaches people how to control their emotions, thoughts and behaviour to improve their mental health.

13

PKU and pregnancy

- Why planning your pregnancy is essential
- Will my children be born with PKU?
- What to expect during pregnancy
- Weight changes during pregnancy
- Exercise during pregnancy
- Obstetric care
- After birth

Why planning your pregnancy is essential

Women with PKU whose blood phe levels are kept low **before** they become pregnant and **throughout their pregnancy** can have normal, healthy babies. Safe phe levels for women planning pregnancy are **lower than usual acceptable levels when not pregnant.** Phe control for pre-conception and during pregnancy aims for levels of 70–250umol/L. Discuss appropriate target levels with your metabolic clinic team.

Many women do not know they are pregnant until several weeks after they have conceived. For babies of women with PKU, high phe levels during these early weeks could cause heart abnormalities and other serious problems, such as low IQ and restricted head growth.

Planning your pregnancy, following a strict diet – before conception so that your blood levels are not high when you become pregnant, and throughout pregnancy – will prevent PKU-related problems. Your metabolic clinic team will help you plan your pregnancy and offer support throughout. They will provide advice on keeping your blood phe levels safe, and advise how often you will be required to do blood tests. Usually this phe monitoring is done at least weekly during pre-conception and throughout the pregnancy.

Becoming pregnant accidentally may cause harm to your baby. If you do fall pregnant unexpectedly, contact your clinic team as soon as possible to discuss your options. Also, if you are not already on diet, go back on it and take the supplement as prescribed. Don't delay!

In Australia and New Zealand all pregnant women and women planning pregnancy are advised to take folic acid. The PKU supplement contains folic acid but it may not meet your needs during pre-conception and pregnancy. Your metabolic clinic team will advise if you need to take additional folic acid or increase the dose of your PKU supplement.

For women who are planning a pregnancy or who are pregnant or breastfeeding, avoiding alcohol is strongly advised.

High levels of phe at any time during the pregnancy may damage your baby.

Will my children be born with PKU?

If you have PKU and your partner does not, the chance of having a baby with PKU is very low – from 1 in 100 to 1 in 120. Diet before or during pregnancy will not alter whether or not your baby will have PKU. PKU is genetic. Refer to 'Genetics and PKU: Frequently asked questions' in chapter 1 for more information.

What to expect during pregnancy

The first 13 weeks

The amount of protein you can eat may not change much during this time. During the early part of your pregnancy, you may feel tired and a little sick. It is important to remember that weight loss during this stage of pregnancy can push your phe levels up. It can be very challenging to maintain enough energy and a very low-protein diet when you're experiencing sickness and have a poor appetite.

If you are feeling nauseated (referred to as 'morning sickness') the following may help:

- small amounts of food very often – eating every hour can help settle your stomach
- don't let your stomach get too empty – have plenty of low-protein and free foods around to snack on. For example, low-protein biscuits, bread, fruit, pasta and cereals
- if you become dehydrated you will feel worse. Drink water, cordial, fruit juice, sports drinks, flat ginger ale or cola (avoid sweeteners containing phe) or eat ice and icy poles/ice blocks
- if you are getting up during the night, have something to drink and eat to stop you from feeling nauseated when you wake up
- having your supplement is important. To make it more palatable try:
  - eating something to settle your stomach 15 minutes before your supplement
  - different flavours, diluting with water, fruit juice or flat lemonade
  - taking smaller amounts more often
- if your nausea continues and you are not able to tolerate your PKU supplement or food, talk to your metabolic team, obstetrician or GP about anti-nausea medications.

Also, during this time:

- commence twice-weekly blood tests for phe – your metabolic clinic will advise how frequently these should be repeated and will contact you if adjustment to your diet is needed
- you may require a supplement of DHA (docosahexanoic acid), an omega-3 fat. Your dietitian will advise if your PKU supplement gives you enough or if extra is needed.

Try to eat regularly to prevent weight loss, as losing even a little weight at this time can make your blood phe level higher.

From 14 to 26 weeks

During this time your phe levels may start to drop as the baby starts to grow much larger – you may be able to start eating more protein in foods. Your dietitian and metabolic team will guide you with frequent contact and instructions on when to increase the protein in your diet. This is based on your regular phe results.

From 27 to 40 weeks

During this time:

- you will continue to increase your daily protein intake – some women with PKU will eat significantly more protein than they did before the pregnancy, and still keep blood phe levels in the target range
- keep fit with regular exercise
- for some women with PKU, it is challenging to include enough protein in the diet at this stage of the pregnancy. Including regular breads, pasta and rice in place of low-protein varieties is an easy way to increase protein intake. Your dietitian can discuss alternatives with you based on your food preferences.

Weight changes during pregnancy

The recommended weight gain during pregnancy for women with PKU is the same as for women who do not have PKU. Recommendations depend on your body mass index (BMI) before pregnancy.

BMI is calculated by:

weight (kg) ÷ [height $(m)^2$]

Weight gain during pregnancy based on BMI*

BMI (kg/m^2)	Weight gain (kg)
<18.4	12.5–18
18.5–24.9	11.5–16
25–29.9	7–11.5
>30	5–9

*Weight gain recommendations are based on a singleton pregnancy. Weight gain for multiple pregnancy should be guided by your doctor. Reference: Guidelines CRIOMPW, Yaktine AL, Rasmussen KM, Board FN, Board on Children YF, Medicine I, et al. Weight Gain During Pregnancy: Re-examining the Guidelines: National Academies Press, 2009.

First 13 weeks

Aim to keep your weight steady, gaining just one to two kilograms in total over the first 13 weeks. Weight gain is supposed to be slow, but weight loss is not good for your phe levels or your baby's growth.

From 14 to 26 weeks

Expect weight gain of around 300–500g each week.

From 27 to 40 weeks

The baby grows rapidly. Weight gain of up to 500 g each week is common.

Exercise during pregnancy

Keeping active during pregnancy helps you to cope with the demands of labour and caring for an infant. It is common to feel tired at certain stages and regular activity is often beneficial.

Your exercise needs will change as your pregnancy progresses. Keep the following in mind:

- if you're unsure about suitable exercise options, discuss it with your doctor, obstetrician or midwife
- include enough energy in the diet to support exercise
- if exercise is causing weight loss, you may need to reduce the amount or intensity for a while and discuss it with your metabolic team.

Obstetric care

Your obstetric care before and during the birth will usually be the same as for a non-PKU pregnancy and birth. The only difference will be that you are on a PKU diet. Your metabolic team will advise your obstetrician, local doctor or midwife about your treatment for PKU during pregnancy. In some cases, you may be referred to a high-risk pregnancy obstetric care service for more detailed ultrasounds and monitoring.

After birth

Like all babies born in Australia and New Zealand, your baby will be offered a newborn screening test to identify the risk of having PKU and other rare conditions. Your metabolic team will advise you and the hospital when the test should be done. It usually occurs 48 to 72 hours after birth.

Breastfeeding is ideal for babies and meets all nutritional needs for the first six months of life. It improves your baby's immune system and reduces risk of allergy and other health issues. Refer to chapter 19 'PKU resources' for breastfeeding support and information. Mothers with PKU can breastfeed and it may keep your phe level lower. Breastfeeding is safe even if your phe is high. It helps your weight (and shape) return to normal. It also means your body needs more protein from food. This will be monitored with fortnightly phe levels and will change over time depending on how much breastmilk your baby drinks.

Staying on diet is strongly recommended after the birth of your baby as high blood phe levels can affect your mood, ability to concentrate and potentially the ability to care for an infant. Keeping your levels lower may help you to create a happier home environment for you and your newborn. If your baby has PKU, you will both need a special diet.

14

Sapropterin (BH4)

- Understanding sapropterin (BH4)
- How does sapropterin work?
- What does ‘sapropterin or BH4 responsive’ mean?
- Starting sapropterin: Setting up for success
- Taking sapropterin
- Balancing nutrition and phe levels
- What is a nutritious diet?
- Unfamiliar foods
- Resources

Understanding sapropterin (BH4)

In some people with PKU, sapropterin can be used as a medication that may help reduce phe levels in the blood. This may increase you or your child's protein tolerance.

Sapropterin may be used on its own, or in combination with a low-protein diet and/or PKU supplements.

In Australia, sapropterin was listed on the pharmaceutical benefits scheme in May 2019 for patients under 18 years old with PKU and some adults previously treated with sapropterin. Discuss individual eligibility and required processes with your metabolic team.

National guidelines for use of sapropterin have been developed. The ASIEM guidelines (2016) provide background:

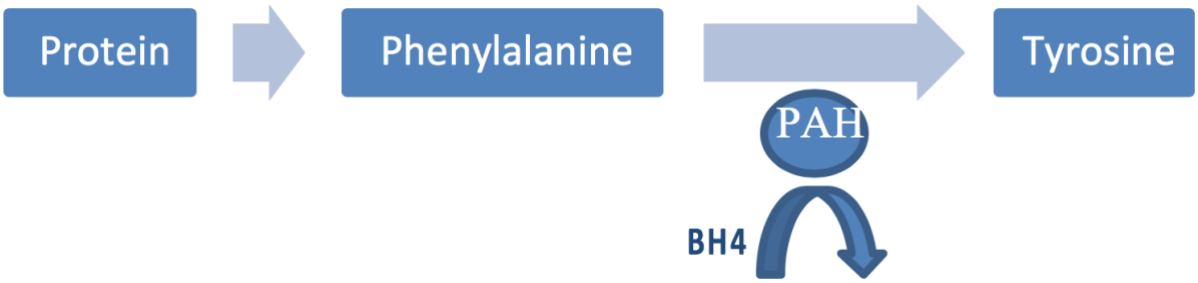
https://www.hgsa.org.au/resources/asiem-resources-for-parents-and-families/australasian-bh4-clinical-guideline

In New Zealand, sapropterin is available for persons with PKU who are pregnant or are actively planning to become pregnant. Sapropterin for use in non-maternal PKU is currently under review. See the link below for more information:

https://www.pharmac.govt.nz/2019/07/01/SA1757.pdf

How does sapropterin work?

The phenyalanine hydroxylase (*PAH*) enzyme needs a 'helper' or coenzyme called sapropterin/BH4. It helps the *PAH* enzyme break down more phe (see chapter 1 'What is PKU?' for more background).



What does 'sapropterin or BH4 responsive' mean?

In some people with PKU, sapropterin significantly boosts the *PAH* enzyme activity, meaning the body can break down much more phe, lowering blood phe levels. These people are known as being 'responsive' to sapropterin/BH4.

In other individuals, the sapropterin may not lower the phe level at all or not enough to be a treatment option for them.

There are different criteria across the world for sapropterin/ BH4 responsiveness.

Discuss your or your child's individual needs with your metabolic team.

To determine if you are responsive, your metabolic clinic team may:

- perform a sapropterin loading dose. This may be done at diagnosis when phe levels are already high. A baby is given a one-off dose of sapropterin before they commence a PKU diet. The change in phe is recorded over 24 hours
- a trial of sapropterin over seven days with regular monitoring of blood phe levels.

In Australia, you are/your child is considered 'sapropterin/BH4 responsive' if:

- there was a ≥30% decrease in phe levels from baseline on responsiveness testing, while maintaining consistent diet +/- PKU supplement intake
- recorded phe levels were over 360 in the first month of life, or over 600 from one month of age and older.

If you/your child are determined to be 'responsive', this medication could be used to assist in managing your PKU. Your metabolic clinic team will guide you through the next stage of commencing regular sapropterin medication and gradually changing over to a higher protein diet and reduced amount of supplement.

Sometimes when phe levels have been stable for a while within the treatment range, you or your child may be able to tolerate more protein. It is important to know, given the high cost of sapropterin, what level of protein can be tolerated with or without this additional medication. Discuss with the dietitian/metabolic clinic team who can plan with you how to gradually increase protein intake prior to using sapropterin.

Sapropterin is not a cure. It is a treatment method that can help control phe levels for some people. Some protein restriction and PKU supplement may still be needed in the diet. While sapropterin may work for a person who is 'responsive', it does not work for everyone.

Starting sapropterin: Setting up for success

Commencing sapropterin and the process of making changes to the PKU diet is not without challenges. If sapropterin is available for you or your child, it's important to consider if the timing is right for your circumstances and if you can meet the extra responsibilities of commencing sapropterin.

Responsibilities of commencing sapropterin

For the transition from a PKU diet to a higher protein diet, responsibilities include:

- increased dry blood spot (DBS) testing. For example, twice weekly, weekly or fortnightly
- regular food diaries and contact with the dietitian
- sapropterin is a medication – you must take it as prescribed every day
- willingness to try higher protein foods and learn about including foods from a variety of food groups.

Questions you could ask yourself or metabolic team before starting sapropterin

- Does my child want to change their PKU diet? Or do I want to change my PKU diet?
- Is my child or am I willing to try new foods?
- Are phe levels well controlled?
- Can I take on the extra responsibilities of trialling sapropterin? Or are there too many other changes in my life right now, such as a new school, exams, a change in exercise or other challenges?
- Can I/my child take medication reliably every day?
- Do I check blood phe levels regularly enough to monitor changes in diet?

Taking sapropterin

The dose of sapropterin may change based on weight or phe levels and it must be taken as prescribed. It can be difficult to remember to take medication every day. Some tips include:

- make it part of your daily routine. Keep it next to your toothbrush or near something you use multiple times a day
- set reminders on your phone or computer
- ask someone to remind you initially
- use a webster/blister pack to portion out the doses for the week
- the tablets can be swallowed whole, or crushed and dissolved in water, or mixed in a small amount of soft food before taking them. Sapropterin powder is also now available.

Balancing nutrition and phe levels

Balancing phe levels and protein, vitamin and mineral intake

Sapropterin can assist the body in processing phe, allowing more protein from food. The amount of protein a person can tolerate is different for everyone. Protein intake targets will be changed gradually based on blood phe levels and discussion with your dietitian.

Target phe levels are not the only important thing in transitioning your diet when on sapropterin.

Initially, the PKU supplement will provide the majority of vitamins and minerals. Over time as supplement intake is reduced, these come from food.

When you or your child's protein allowance goes up, it may be tempting to eat more foods that have been limited in the past such as hot chips, chocolate and cake. It's important to eat a balanced diet so you or your child gets all the protein, vitamins and minerals (such as iron and calcium) needed for a healthy body and mind. If protein intake is too low or consumed from vegetable-based foods only, phe levels may be too low for growth and rebuilding of tissues such as muscle. There is also a higher risk of vitamin and mineral deficiencies.

What is a nutritious diet?

Your dietitian will provide individual advice to ensure you are meeting your nutritional needs while not exceeding phe target levels when taking sapropterin. An example of a nutritious diet is choosing a variety of foods from the five food groups:

- vegetables
- fruit
- lean meats, poultry, fish, eggs, tofu, nuts and seeds, legumes/beans
- grain (cereal) foods, mostly wholegrain and/or high fibre varieties
- milk, yoghurt, cheese and/or alternatives, mostly reduced fat.

Protein tolerance varies when taking sapropterin. Not everyone will be able to tolerate high-protein foods, whereas others will be able to eat from all five food groups.

Some foods are not part of the five food groups because they are high in either calories (energy), saturated fat, sugar, salt or alcohol, and have low levels of important nutrients. These foods should only be eaten occasionally.

Sapropterin may allow relaxation but not necessarily cessation of the PKU diet or supplements. In Australia, once you are consuming regular carbohydrates (regular bread, rice or pasta) the low-protein food grant is no longer available.

Unfamiliar foods

Trying new foods can be challenging. Unfamiliar foods such as meat and dairy may have an unusual texture or flavour. They are valuable sources of vitamins and minerals and should be introduced to the diet and eaten regularly, depending on your protein allowance. You will be guided by your metabolic clinic.

Some tips to get started if you are able to tolerate higher protein foods in your diet

- It takes time to adapt to new flavours and textures. Repeated exposure to new foods over time without pressure to eat assists with familiarity.
- Let your child choose what meat/alternatives/dairy option they would like to try.
- Start small. Include small pieces in a dish with familiar foods such as soup, stir fries or curries. It makes it less challenging or overwhelming.
- Trial slow-cooked, soft or minced meat mixed through familiar rice or vegetable dishes.
- Thinly sliced roast meat and soft meats such as white fish or slow-cooked beef can be a good starting point. It’s easier to learn to chew soft meat rather than a steak.
- Try different types of the same food. For example, different types of yoghurt, custard, cheese or meat, chicken, fish or eggs prepared in different ways.

Resources

The following resources provide more information on introducing new foods and expanding diet variety and nutrition.

https://www.ellynsatterinstitute.org/resources-and-links-for-the-public/

Australian Government websites

https://www.healthykids.nsw.gov.au/

https://www.makehealthynormal.nsw.gov.au/home

http://raisingchildren.net.au/healthy_eating/school_age_healthy_eating.html

https://www.betterhealth.vic.gov.au/health/healthyliving/food-variety-and-a-healthy-diet

https://www.eatforhealth.gov.au/

Children's hospital factsheets on healthy eating

https://www.schn.health.nsw.gov.au/fact-sheets/category/#cat23

https://www.rch.org.au/nutrition/resources/

15

Dental care and PKU

- Tooth decay and dental erosion
- Why are children with PKU at greater risk of dental decay and erosion?
- How can dental disease be prevented?

Tooth decay and dental erosion

Tooth decay and dental erosion can affect the teeth of any child. Children with PKU are at greater risk, and adults with PKU also need to take good care of their teeth.

Tooth decay occurs when bacteria in the mouth change sugars in food and drinks to weak acids which eat away the teeth to form holes. Dental erosion occurs when strong acids in foods, drinks or supplements dissolve away the outer layers of the teeth.

Why are children with PKU at greater risk of dental decay and erosion?

Sugary foods and acidic drinks may be consumed more often

The diet for children with PKU includes little protein from foods, so they need to have more carbohydrates and fat to give them enough energy. Carbohydrates, particularly when they're in sugars, biscuits, jam, honey, cordials, juices, fruit drinks, sports drinks, soft drinks, lollies and some snack bars, are more likely to cause tooth decay when consumed often through the day.

They may snack more frequently

The more often and the longer acidic foods or drinks are in contact with the teeth, the more likely it is that surface erosion will occur.

Phe-free supplements are both acidic and sweetened. They are best consumed within 10 to 15 minutes rather than sipped over long periods. Constantly sipping sweetened or acidic fluids increases risk of dental disease.

How can dental disease be prevented?

By toughening the teeth with fluoride

Generally, it is recommended that:

- a child-strength fluoride toothpaste be used with children from 18 months of age and an adult-strength toothpaste from six years. However, because children with PKU may be at higher risk of decay, a dentist may recommend using an adult-strength toothpaste before six years. It is important to discuss this with your regular dentist
- adult supervision of children when brushing their teeth is essential to ensure the correct amount of toothpaste is used and the brushing is effective. Dentists recommend a pea-sized amount of toothpaste for children and adults
- children should brush at least twice daily – especially after breakfast and last thing at night (avoid snacks, supplement and fluids other than water afterwards)
- adults should use a full-strength fluoride toothpaste twice a day – after breakfast and before bed
- after brushing spit out excess toothpaste but don't rinse.

Give the teeth a rest from food and drinks:

- only put infant formula or water in a baby's bottle
- remove the bottle after each feed and do not put your baby to sleep with a bottle
- move to an open cup by the age of 12 months
- drink larger amounts over a short time frame rather than sipping a drink slowly
- give teeth a rest – aim for three meals a day and minimise snacks between meals
- avoid giving sweet food or drink at bedtime
- water is the best drink to have apart from the supplement – give some after each supplement drink and often during the day
- keep soft drinks, cordials and juice for special occasions.

Have regular check-ups with a dental practitioner:

- early visits help your child get used to seeing a dentist – this can make it a lot easier as they get older
- the dentist needs to know that you or your child have PKU and to understand what it is
- ask your dentist for preventative advice and treatment
- your dentist will be able to pick up signs of decay and erosion much earlier than you or your doctor will.

SUGAR OR SUGAR-FREE?

Sugar is useful in the PKU diet as it helps to provide energy. Sugar-free foods don't do this and may provide extra phe (see 'Artificial sweeteners' in chapter 6). The important thing when eating sugary food is to avoid constant snacking or sipping, give teeth a rest and maintain good dental care.

16

Sick days

- What happens to phe levels during illness?
- What to do if your baby is sick
- Feeding your sick baby
- If your baby has gastroenteritis
- What to do if you or your child are sick
- Gastroenteritis
- Medications containing phe

IMPORTANT NOTE:

Contact your local doctor if you, your baby or your child are unwell just as you would if you/they didn't have PKU. They will assess hydration and provide specific advice. The information below is a guide but does not replace medical advice.

What happens to phe levels during illness?

Illness can cause a temporary rise in your phe levels, but no long-term harm. When you are unwell the body starts to break down its own tissues, releasing phe into the bloodstream.

What to do if your baby is sick

Infections and other illnesses frequently affect babies and young children, and those with PKU are no different.

Feeding your sick baby

Food and fluid intake are often less than usual when your baby is sick. Maintaining enough fluid intake to ensure your baby is hydrated is important. Here are some practical guidelines (not relevant if your baby has gastroenteritis symptoms such as vomiting and diarrhoea):

- offer small feeds more frequently than usual
- if your baby has a poor appetite and is feeding poorly from the breast you may have to express to keep up your supply
- your dietitian may recommend you increase your baby's energy intake and will explain how to do this.

If your baby has gastroenteritis

See your doctor if your baby has gastroenteritis as it is important to avoid dehydration.

What to do if you or your child are sick

If you or your child are unwell:

- encourage adequate fluid intake by sipping on/offering drinks every hour or two while you are/your child is awake
- dilute fluids and supplement – this may make it easier to drink them
- don't force-feed your child – they may reject the supplement later
- eat/offer foods according to appetite. It is ok if the full protein allowance is not consumed. Return to usual protein intake as appetite returns
- plain foods such as low-protein toast, rice or pasta and hydrating foods such as watermelon or ice blocks are easier to tolerate.

Gastroenteritis: Guidelines for children and adults

These general guidelines are for children and adults. They are not appropriate for babies:

- for the first 12–24 hours offer mainly oral rehydration solution (ORS) – see further information on the following page – or clear fluids such as water and very dilute cordial. For example, offer 100–200 ml every hour or two when your child is awake
- then reintroduce the supplement and usual diet with the addition of ORS or clear fluids – the supplement may be better tolerated if it is more dilute
- it is not necessary to consume all the supplement or to have the full amount of protein intake until the appetite returns.

Medications containing phe

All medications prescribed by your doctor are suitable, but if possible, avoid those with the artificial sweeteners aspartame, NutraSweet or additives 951 or 962. If a medication is required urgently and contains these sweeteners, give it until you can get an alternative or discuss it with your metabolic team doctor or nurse practitioner. Over-the-counter medicines should also be checked for artificial sweeteners.

Oral rehydration solution (ORS)

A number of oral rehydration solutions are available from your pharmacy. These contain electrolytes and sugar. Some are sweetened with aspartame, NutraSweet, or additives numbers 951 and 962 and are not suitable for people with PKU. Use them only in an emergency until an alternative is available. Your pharmacist can advise about other suitable products. Always check the label.

If commercial ORS is not available, or it is not your/your child's preferred option, make up one of the following:

- dilute cordial: use 10 ml + 150 ml water
- dilute soft drink or lemonade: use 50 ml + 150 ml water
- dilute fruit juices or fruit drinks: use 50 ml + 150 ml water
- sugar water: use 1 level 5 ml teaspoon sugar + 250 ml water.

Do not use phe-containing sugar-free or low joule or 'diet' cordials or soft drinks.

17

Overseas travel

- Covering letter
- Sealed containers
- Organising supplies for your trip
- Lost luggage
- Food and drink for the flight
- Travellers' tips
- Checklist

There is no reason to let PKU stop you from travelling. Holidays and travel away from home, particularly if you're going overseas, requires careful planning. As you get used to the experience of being away from home and maintaining the diet, your confidence will increase.

Covering letter

Always take a customs declaration letter from your metabolic clinic. Let your metabolic clinic know in advance that you are planning to travel to allow them enough time to prepare your travel letter.

The covering letter should list the name and quantity of all the PKU products you will be carrying in your luggage, stating who they are for and why they are needed. Unless it is officially explained that these products are medical necessities, there may be problems taking them into another country. It is a good idea to make several copies of this letter so you can keep one in your hand luggage, and one in your checked luggage. You could also take a photo of the document using your mobile phone.

Carrying PBS-listed products from Australia and PHARMAC-listed products from NZ on behalf of someone who is not travelling with you is not allowed.

Sealed containers

Keep the formula or supplement in its ***original sealed container*** and take a copy of the prescription with you. Ensure the pharmacy label is visible on the supplement packaging, as you must show where the medicine was dispensed, the name of the person it was prescribed for and the cost.

If you intend to carry large amounts of products out of Australia it is also wise to complete an ***Export Medication Declaration***, listing supplement, low-protein products and prescribed medications. This form is available from doctors, nurse practitioners, pharmacists or online at:

https://www.humanservices.gov.au/sites/default/files/documents/2619-1201en.pdf

Organising supplies for your trip

For longer periods of travel, you will need an additional script from your metabolic team so you can obtain extra supplies of supplement and special foods before you leave.

Lost luggage

It's a good idea to carry extra supplement to cover the possibility of your luggage being lost. Divide it between your suitcases and hand luggage. Make sure you have enough supplement in your hand luggage to last a few days in case you arrive at your destination before your checked luggage.

Food and drink for the flight

Contact the airlines beforehand to organise food that you, or your child, can eat during the trip. It may be easier to tell them what you *can* eat, rather than what you can't. Take plenty of snacks and supplement for the flight and to cover the possibility of travel delays.

Bottled water

Use uncarbonated bottled water to make powdered formula or supplement. As long as the bottle is sealed, you will have a clean, cold source of liquid which will minimise the chance of getting an infection. For babies, this water should be boiled then cooled. Remember to pack a formula shaker or mixer.

Do not mix your supplement with water until you are past the security checkpoint. You may be asked to discard it due to fluid restrictions when clearing security for international flights.

Ready-to-drink supplements can be easier when travelling but are often heavier to carry and may not be permitted in carry-on luggage when flying. Ask your metabolic dietitian for suitable options.

Travellers' tips

The following tips from people with PKU and their families may be helpful:

- choose accommodation with access to kitchen facilities so you have the option to make low-protein meals
- you may be able to freight the supplement to your destination beforehand to reduce your luggage, or request a powder PKU supplement which is easier to pack and lighter to carry
- some post the supplement to themselves at poste restante (the local post office in a particular city) or to their hotel and collect it when they arrive. If you're posting supplement (or formula) overseas, include in the package a copy of the doctor's letter and local contact numbers. Do this well ahead to allow for delays
- ask your metabolic clinic team (well in advance of your trip) for information on where low-protein supplies can be obtained and whether there are any reciprocal agreements about prescriptions in the countries you are visiting
- take a dictionary or use a mobile phone app to help with translation
- put together a list of foods you can eat. If you're travelling on an organised tour, the travel company may be able to have it translated or you can have it translated online so you can use it when visiting restaurants
- arrange to have someone at home on standby to post any items you may have forgotten
- before you leave, search the internet about PKU associations in the country/countries you intend to visit.

Checklist

Have you packed:

- formula or supplement
- mixing shaker, bottle or drink container
- scoop
- vitamin and mineral supplements if needed
- prescription
- low-protein foods
- customs letter
- scales, if needed
- diet information
- food for your trip.

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Financial assistance

- New Zealand
- Australia
- Government allowances

New Zealand

Some low-protein products are available on prescription and are fully funded. Other products can be purchased directly from the supplier. Contact details/order forms for these products are available from your metabolic dietitian.

Australia

Specialised low-protein products can be purchased directly from the suppliers. Contact details/order forms for these products are available from your metabolic dietitian or nurse.

Government allowances

New Zealand

Ask your doctor or metabolic nurse for advice on financial support that you may be eligible for. Or contact Work and Income New Zealand: phone 0800 559 009. Or visit:

www.workandincome.govt.nz

The usual prescription charges apply when obtaining PKU supplements and low-protein foods. Your child will be eligible for the Child Disability Allowance. A referral to a social worker may be made for further financial assistance if required.

Australia

Government allowances and the rules regarding financial support may change over time. Your social worker or Centrelink will be able to give you up-to-date information. For children, you can apply for three types of financial support: the Health Care Card, the Carer's Allowance and the Inborn Errors of Metabolism (IEM) allowance. Adults are eligible for the IEM allowance.

The Health Care Card

People with PKU may be eligible for the Health Care Card which means they are entitled to prescription medicines, such as PKU supplements, at the lower (pensioner) rate. Eligibility is means tested. Visit:

https://www.humanservices.gov.au/individuals/services/centrelink/health-care-card

The Carer's Allowance

Parents of children with PKU automatically receive the Carer's Allowance, which is paid through Centrelink. Parents must be Australian residents and eligibility is means tested. Visit:

https://www.humanservices.gov.au/individuals/services/centrelink/carer-allowance

or call 132 717

The applications for the Health Care Card and Carer's Allowance are made via Centrelink. Included in the application is a medical report form, which needs to be completed by your child's doctor.

The Inborn Errors of Metabolism Allowance

People with PKU who are on a low-protein diet are entitled to claim an allowance through the Commonwealth Department of Health. This allowance, for individuals with an IEM, is to help offset their food costs. To qualify for the allowance the person must be an Australian resident, older than six months, on an appropriate diet for their PKU, submitting regular blood tests and attending clinic.

It is at the discretion of your clinical team if this form is signed off based on your adherence to your PKU diet, supplements, blood test results and clinic attendance. The allowance is currently paid monthly. This allowance is exempt from a means or assets test. Claim forms are available online and need to be signed by your doctor every 12 months. Visit:

https://www.health.gov.au/internet/main/publishing.nsf/Content/Inborn+Error+of+Metabolism+Programme#Contact

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PKU resources

- PKU associations in New Zealand
- PKU associations in Australia
- Breastfeeding support and information in New Zealand
- Breastfeeding support and information in Australia
- PKU supplement, low-protein food and recipe websites
- Overseas associations
- Sample letters

PKU associations in New Zealand

Contact the National Metabolic Service for information regarding support for PKU in New Zealand.

https://www.starship.org.nz/directory-of-services/metabolic-service

PKU associations in Australia

Metabolic Dietary Disorders Association (MDDA)

MDDA is a national association for people with a variety of metabolic disorders including PKU. It provides support, social activities, conferences, recipe ideas and a quarterly newsletter. They play an important role in advocating for improved patient care and medications and lobby to the government to assist with policies such as the IEM food grant.

Contact: Croydon office:

Shop 4, 6 Thomas Brew Lane, Croydon VIC 3136

PO Box 33, Montrose VIC 3765

Phone: (03) 9723 0600

Freecall: 1800 288 460

Email: office@mdda.org.au

Website: www.mdda.org.au

Facebook: https://www.facebook.com/metabolicdietarydisordersassociation/

The PKU Association of NSW

This support association for families with PKU also has members across Australia and overseas. It publishes a quarterly newsletter with information, updates and recipes. It also organises social events, including an annual Christmas party and a PKU Youth Camp every two years, and raises funds for PKU research. The association is run by adults with PKU and family members of children with PKU.

Contact:

PO Box 4384

Castlecrag NSW 2068

Email: info@pkunsw.org.au

Website: www.pkunsw.org.au

Facebook: https://www.facebook.com/PKU-Association-of-NSW-169183236472835/

Breastfeeding support and information in New Zealand

Practical breastfeeding support is available from your midwife or Plunket Nurse.

Plunketline offers free telephone support 24 hours day, seven days a week on 0800 933 922.

La Leche League

This organisation offers telephone help at any time and has local support groups that meet regularly. Website https://lalecheleague.org.nz/

Breastfeeding support and information in Australia

Breastfeeding support and advice are available from your midwife, maternal and child nurse or lactation counsellor.

Australian Breastfeeding Association

Telephone counselling is available seven days a week via the national breastfeeding helpline. It is staffed by trained, volunteer counsellors.

Contact: 1800 mum 2 mum (1800 686 268)

Website: www.breastfeeding.asn.au

Raising Children website

Provides free, reliable, up-to-date and independent information. Topics include different ages and stages, from nurturing a newborn to raising a confident, resilient teen, and self-care for parents.

Website: https://raisingchildren.net.au/

Australian Government Inborn Errors of Metabolism Program

Provides information on the Inborn Errors of Metabolism Program, including the IEM food grant and application form.

Contact:

IEM program officer, Portfolio Strategies Division Department of Health

MDP 1060, GPO Box 9848

CANBERRA ACT 2601

Phone: (02) 6289 8980

Email: iemprogramofficer@health.gov.au

Website:
https://www.health.gov.au/internet/main/publishing.nsf/Content/Inborn+Error+of+Metabolism+Programme#Contact

PKU supplement, low-protein food and recipe websites

The following websites provide information on PKU supplements, low-protein foods and/or recipe ideas. Your dietitian can discuss suitable supplements for you and arrange samples. Food items on listed websites may not be available for purchase from NZ.

Modify the recipes according to your/your child's protein allowance. Many regular recipes can be adapted so they are suitable for a low-protein diet. Your dietitian can assist with this.

Cortex Health

Available for purchase: low-protein foods.

Home delivery service for PBS-listed PKU supplements in Australia.

Provides recipe ideas.

Website: www.cortexhealth.com.au

MDDA

Provides recipe ideas.

Website: www.mdda.org.au

Nutricia

Available for purchase: low-protein foods.

Home delivery service for PBS-listed PKU supplements in Australia.

Provides recipe ideas.

Website www.pkuconnect.com.au/

NZ website: https://www.pkuconnect.com.au/nz/

Orpharma/Mevalia

Available for purchase: low-protein foods.

Home delivery service for PBS-listed PKU supplements.

Australian website: http://www.orpharma.com/products/low-protein-food/

New Zealand website: https://mevalia.co.nz/

Platypus Foods

Available for purchase: low-protein foods.

Provides recipe ideas.

Website http://www.platypusfoods.com.au

Vitaflo

Available for purchase: low-protein foods.

Home delivery service for PBS-listed PKU supplements in Australia.

Website: www.vitaflo.com.au

Overseas associations

UK PKU association

www.nspku.org

American PKU Associations

www.pkunews.org

www.pkuil.org

www.pkunetwork.org

Sample letters

Your metabolic team can provide resources and letters which can be given to teachers, parents of children in your child's class, carers and health professionals who need to know about your child's PKU.

Glossary

Amino acid

Amino acids are the basic building blocks of proteins. The body makes many amino acids, and others must be obtained from food.

Aspartame

This is an artificial sweetener which contains phe and should be avoided by people with PKU. It may be listed on food labels as NutraSweet, Equal, Canderal, aspartame-acesulphame or additive 951 and 962.

BH4

BH4 is short for tetrahydrobiopterin. PKU is caused by a deficiency in the activity of *PAH*. For *PAH* to function properly, BH4 is needed. In some people with PKU, taking BH4 supplements, known as sapropterin, may improve the function of *PAH*.

Calorie

A calorie or kilojoule is a measure of energy.

Carbohydrate

One of the three main nutrients in food. Foods that provide carbohydrate are bread, pasta, rice, vegetables, fruits and sugars.

Coenzyme

A substance that enhances the action of an enzyme (see enzyme for more information).

Energy

Energy is the capacity of the body to do work. The body derives its energy from the carbohydrate, fat and protein in food. A kilojoule or calorie is a measure of energy.

Enzyme

An enzyme is a protein that facilitates a specific chemical reaction. Enzymes are sometimes described as helpers.

Essential amino acid

An essential amino acid cannot be made by the body and must be obtained from the diet. Phe is an essential amino acid.

Fat

One of three main macronutrients that supply energy to the body. Fat is a high energy food source.

Folic acid

Also called folate. One of the B vitamins. It is recommended for all pregnant women to help prevent birth defects.

Gene

A unit of heredity found in all cells in the body. Genes carry hereditary information for bodily processes and traits, such as blood group and hair colour, and instructions for producing chemicals.

Gram

One thousandth of a kilogram. A system of counting grams of protein is used to keep track of phe intake. In most foods, one gram of protein contains 50 mg of phe.

Hormone

A chemical made by the body. Hormones circulate in the blood and control the actions of certain cells or organs.

Kilojoule

A kilojoule or calorie is a measure of energy.

Metabolic clinic team

Metabolic clinic teams vary depending on where you live. Team members may include a doctor, dietitian, nurse, laboratory scientist, newborn screening biochemist and a social worker or psychologist.

Newborn screening

All newborn babies in Australia and New Zealand have a blood test within the first few days of birth to screen for conditions such as PKU.

PAH

The abbreviation for phe hydroxylase.

Phe

The abbreviation for phenylalanine. An essential amino acid found in protein foods. It is normally converted to tyrosine in the body. It also plays an important role in the formation of brain chemicals or neurotransmitters. People with PKU cannot break down phe in foods in the usual way.

Phe hydroxylase

An enzyme needed by the body to break down the phe in protein foods. People with PKU do not have this enzyme in sufficient amounts, so they cannot break down phe to tyrosine.

Phenylketonuria

An inherited condition where the body lacks the enzyme phenyalanine hydroxylase needed to break down phe. The condition is treated from soon after birth with a special diet. Following treatment for life is recommended.

PKU

See phenylketonuria.

PKU formula

Also called the supplement. This is taken by people with PKU to replace the protein in their diet. It contains all essential amino acids (except phe or containing negligible amounts of phe), plus vitamins, minerals and extra tyrosine. It is vital for people with PKU.

Plunket Nurse

The New Zealand equivalent of Australia's maternal and child health nurse. They provide support from birth to five years of age.

Protein

Protein is made up of amino acids. It is needed by the body for growth and repair. Many foods contain protein. Foods such as meat, fish, eggs, milk, nuts, soy and legumes are rich in protein and are not suitable for people with PKU. A protein supplement makes up for the protein they are unable to eat in food.

Micromoles per litre (µmol/L)

A unit of measurement used to describe the amount of phe in the blood.

Protein supplement

See supplement.

Supplement

Also called PKU formula. This is given to people with PKU to replace the protein in their diet. It contains all essential amino acids (except phe or containing negligible amounts of phe), plus vitamins, minerals and extra tyrosine. It is an essential part of the treatment for PKU.

Tyrosine

Tyrosine is an amino acid that is partly obtained from phe. Used by the body to produce hormones, skin and hair pigment, it is also considered vital to normal mental functioning. People with PKU cannot convert phe to tyrosine. They obtain tyrosine from the supplement.

PROTEIN COUNTED & FREE

Vegetables & Fruit in PKU

Fruits and vegetables are healthy and low in protein, making them an important part of the PKU diet. These lists, which are based on phenylalanine content, should be used for counting fruit and vegetables. If a fruit/vegetable comes in packaging with a nutrition information panel (NIP) the following rules apply:

- If it contains only free (uncounted) fruit/vegetable e.g. canned tomatoes with basil, do not count these foods
- If the fruit/vegetable is combined with other foods which you would normally count e.g. with flour in a fruit bar use the value as per the packaging NIP.

Fresh Fruit Counting

Most fresh, frozen & canned fruits do not need to be counted on a low protein diet. Only a small number of fruits need to be counted if eaten in larger amounts. The weight of edible fruit equivalent to <u>one gram</u> (1g) of protein is listed below.

BANANA

65g

MULBERRY

75g

PASSIONFRUIT PULP

40g

POMEGRANATE SEEDS

85g

Dried Fruit Counting

Dried fruits generally contain more phenylalanine than fresh, frozen or canned fruits. If you eat large amounts of any dried fruit not listed below talk to your metabolic dietitian as it may need to be counted. Below, the weight equivalent to <u>one gram</u> (1g) of protein in each dried fruit is listed.

APRICOTS

55g

FIGS

60g

GOJI BERRIES

20g

SULTANAS

85g

Please note that images pictured do not reflect the weights listed

Vegetable Counting

The weights listed for each of the vegetables below is equivalent to **one gram** (1g) of protein.

ASPARAGUS

65g

AVOCADO

90g

BEAN SPROUTS (MUNG)

60g

BROCCOLI

30g

BROCCOLINI

30g

BRUSSEL'S SPROUTS

65g

CAULIFLOWER

70g

CORN KERNELS

35g

CORN ON COB

4cm

KALE

35g

MUSHROOMS

60g

PEAS

25g

Please note that images pictured do not reflect the weights listed

Vegetable Counting Continued

The weights listed for each of the vegetables below is equivalent to <u>one gram</u> (1g) of protein.

PUMPKIN
(EXCEPT JARRAHDALE)

65g

ROCKET

35g

SILVERBEET

45g

SNOW PEAS

55g

SPINACH
(FRESH)

45g

SPINACH
(BOILED / FROZEN)

45g

TARO

70g

YAM

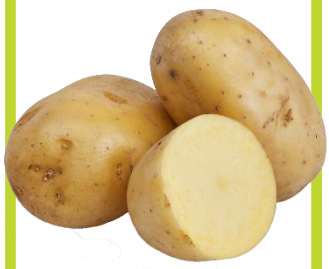
70g

Please note that images pictured do not reflect the weights listed

Potatoes

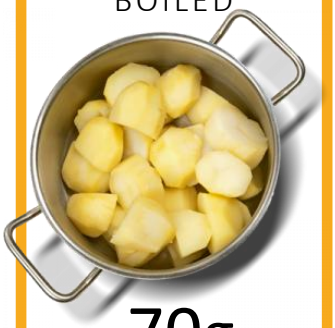
The phenylalanine content of potatoes depends on the cooking method. The weight of potato equivalent to **one gram** (1g) of protein is listed below.

RAW



50g

BOILED



70g

ROASTED

40g

CHIPS

40g

The weight of sweet potato equivalent to **one gram** (1g) of protein is listed below.

RAW

45g

BOILED

45g

ROASTED

45g

CHIPS

45g

Please note that images pictured do not reflect the weights listed

Examples

Now that you know the weight of various fruits and vegetables equal to 1g of protein you can use a calculator and kitchen scales to determine how much protein is in your portion size with this calculation:

Weight of fruit or vegetable	÷	Weight equal to 1g protein	=	Grams of protein

Example 1: Roasted Sweet Potatoes

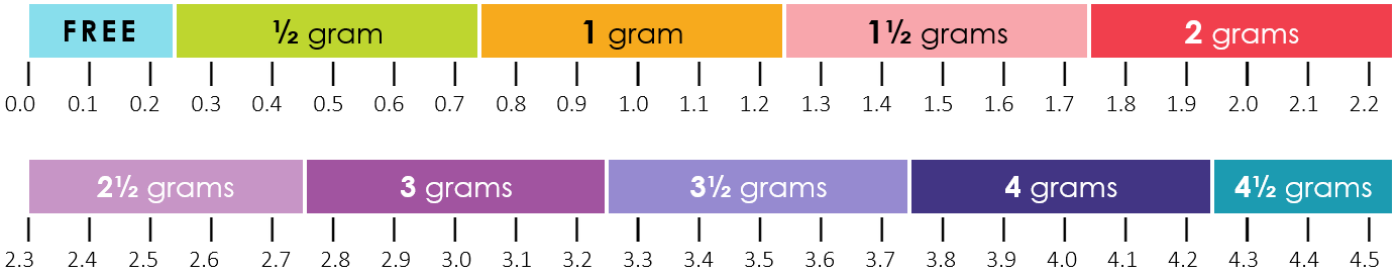
STEP 1: Place a plate or bowl on your kitchen scales and set them to zero (or TARE).

STEP 2: Place the amount of roasted sweet potatoes you intend to eat on the scales. In this case it is 157g.

STEP 3: Use the calculation:

Weight of vegetables (g)	÷	Weight equal to 1g protein	=	Grams of Protein
157		ROASTED  45g		3.4889

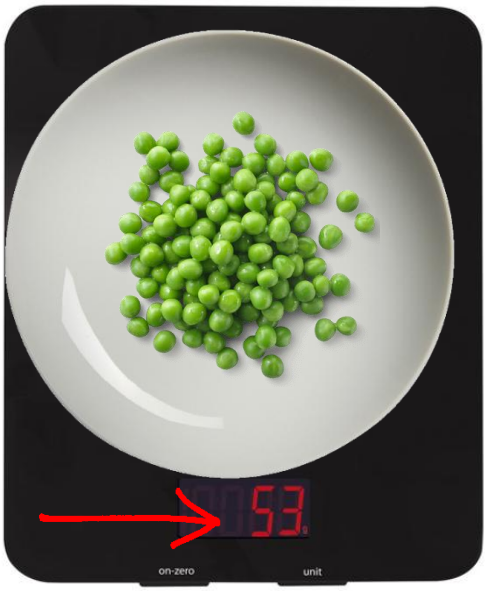
STEP 4: Round to the nearest half gram of protein. If you find this difficult use scale below (you only need to use the first number after the dot, ignore the others). In this example 157g roast sweet potatoes contains 3½ grams (3.5g) of protein.



Examples

Weight of fruit or vegetable	÷	Weight equal to 1g protein	=	Grams of protein

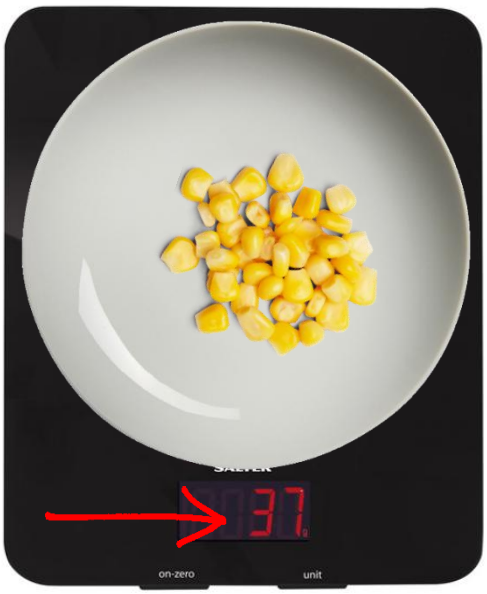
Example 2: Peas



Weight of vegetables (g)	÷	Weight equal to 1g protein	=	Grams of protein
53	÷	PEAS  25g	=	

In this example 53g of peas contains 2 grams (2g) of protein when rounded to the nearest half.

Example 3: Corn kernels

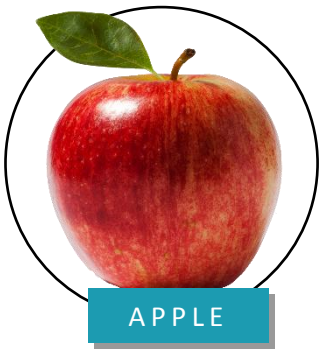


Weight of vegetables (g)	÷	Weight equal to 1g protein	=	Grams of protein
37	÷	CORN KERNELS  35g	=	

In this example 37g of corn kernels contains 1 gram (1g) of protein when rounded to the nearest half.

Protein Free Fruit List

The fruits and vegetables in the following lists do not need to be counted in the PKU diet if standard portion sizes are used. Although these foods are classified as “protein free” they still contain small amounts of phenylalanine. If you eat large portions of any of these foods talk to your dietitian about whether this should be counted.

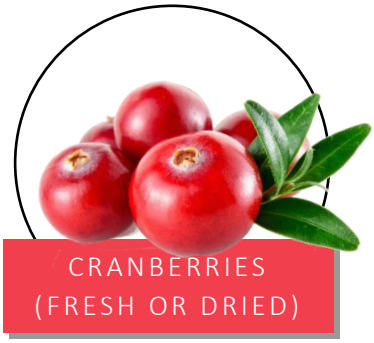

APPLE


APRICOT (FRESH ONLY)


BLACKBERRIES


BLUEBERRIES


CHERRIES


CRANBERRIES (FRESH OR DRIED)


CUMQUAT


CURRANTS (FRESH OR DRIED)


CUSTARD APPLE

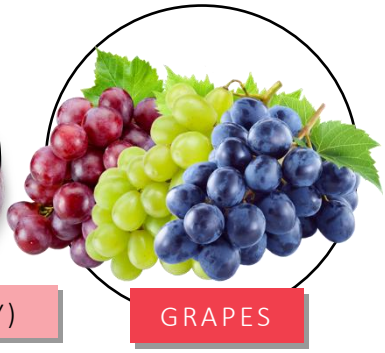

DATES


DRAGON FRUIT


FEIJOA


DURIAN


FIGS (FRESH ONLY)

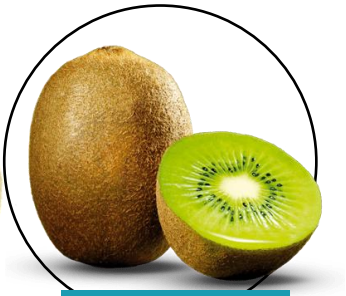

GRAPES


GRAPEFRUIT

Protein Free Fruit List


GUAVA


HONEYDEW MELON


KIWI FRUIT


JACK FRUIT


LEMON & LIME


LONGAN


LOQUAT


LYCHEE


MANDARIN


MANGO

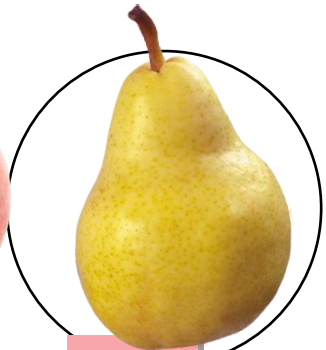

MANGOSTEEN


NECTARINE


ORANGE


PAWPAW (PAPAYA)

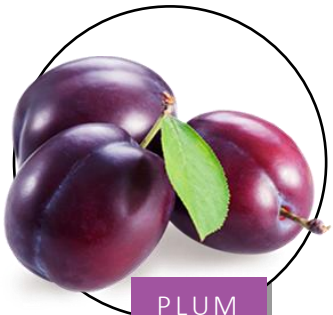

PEACH


PEAR

Protein Free Fruit List


PERSIMMON


PINEAPPLE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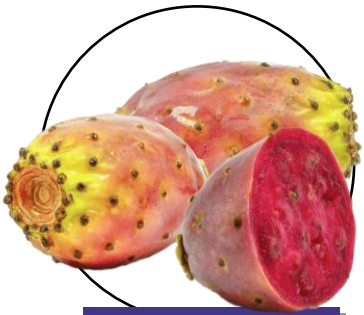
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PRUNES


QUINCE


RAISINS

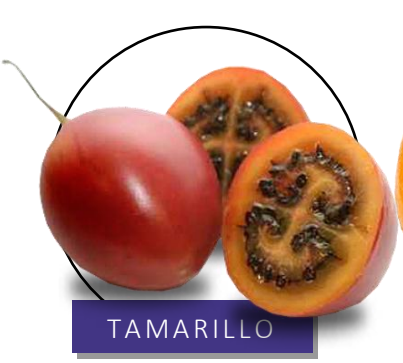

RAMBUTAN


RASPBERRY


ROCKMELON


STAR FRUIT


STRAWBERRY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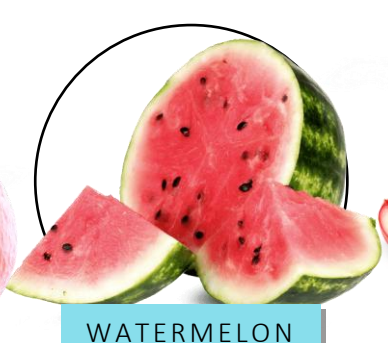
TANGELO & TANGARINE


WATERMELON


WAX JAMBU

Protein Free Vegetable List


ARTICHOKE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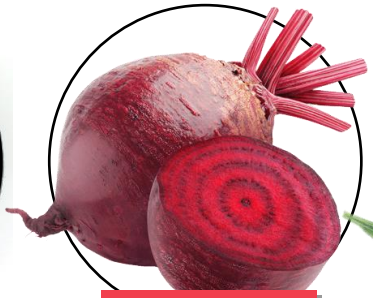

BEETROOT

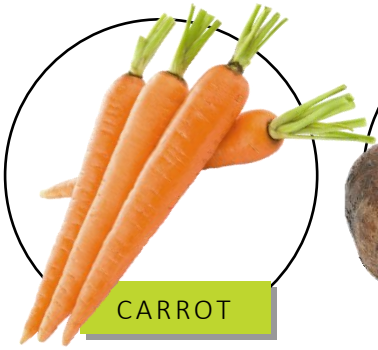

BITTER MELON


BOK CHOY


CABBAGE


CAPERS


CAPSICUM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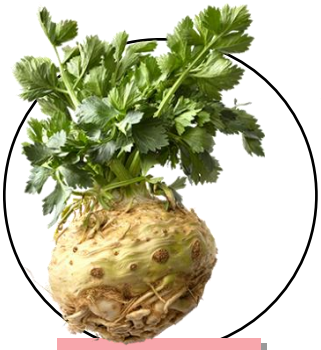
CASSAVA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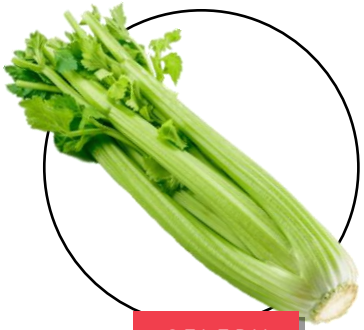
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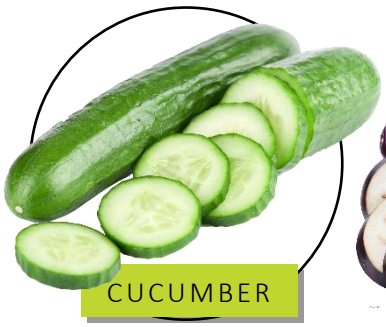
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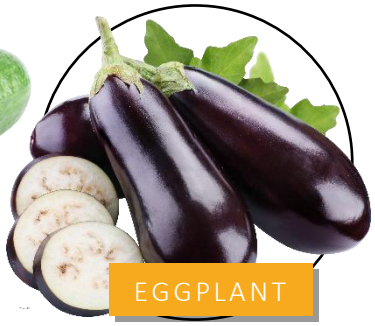

EGGPLANT


ENDIVE

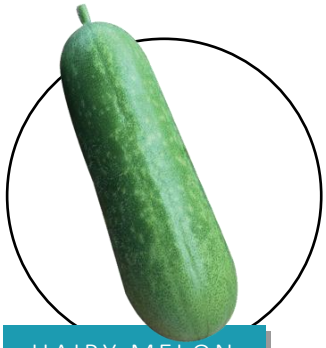

FENNEL


GHERKIN

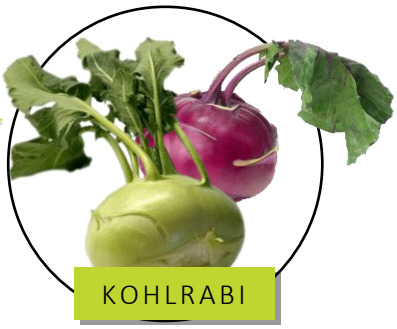

GARLIC & GINGER

Protein Free Vegetable List


GREEN BEANS


HAIRY MELON


HERBS & CHILLI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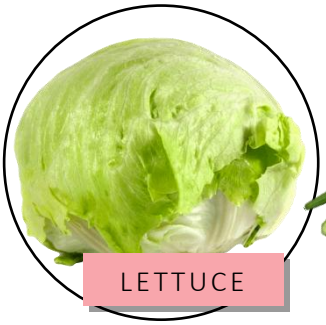
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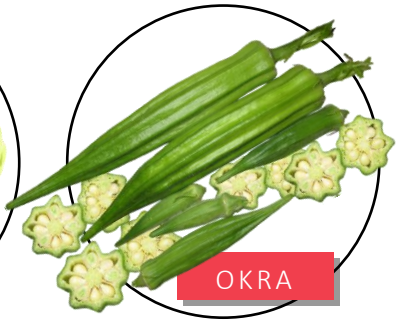
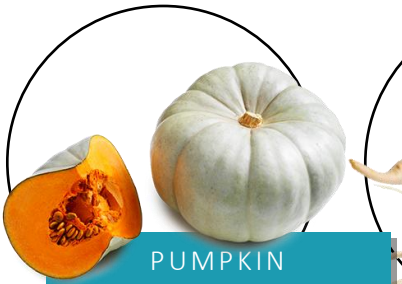

OKRA


OLIVES

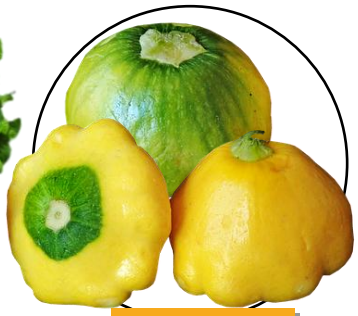

ONIONS, ESCHALLOTS & SPRING ONIONS


PARSNIP


PUMPKIN (JARRAHDALE ONLY)


RADISH


RHUBARB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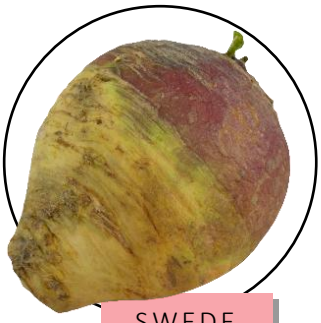
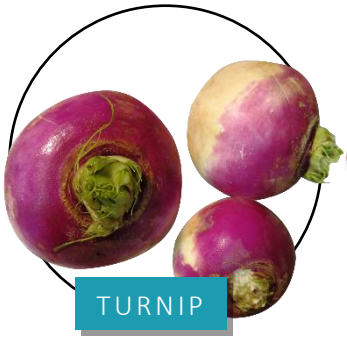

SWEDE


TOMATO


TURNIP


WATER CHESTNUT


WATERCRESS


ZUCCHINI

Label Reading

ON A LOW PROTEIN DIET

Label reading is an essential skill for following a low protein diet. Understanding how to read the nutrition information panel on the label will help you to make smart decisions about what to eat without going over your protein allowance. Don't worry if you find label reading confusing at first - many people do! After reading this guide you will have the following skills to count protein like an expert.

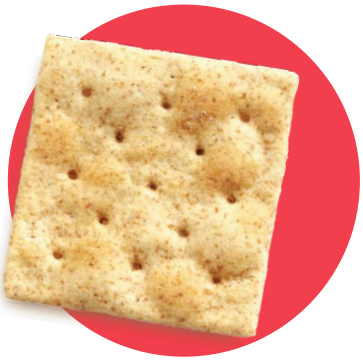
- ☑ Identify the "per serving" and "per 100g" column on the nutrition information panel and decide which one to use
- ☑ Understand what the nutrition information panel classifies as a "serving"
- ☑ Calculate protein "per serving" to the nearest ½ gram (0.5g)
- ☑ Calculate the weight of any food equal to one gram of protein using the "per 100g" column
- ☑ Calculate how many grams of protein in the weight of any food using the "per 100g" column
- ☑ Identify and avoid foods containing aspartame

Using the "per serving" column

The "per serving" column can be used when the serving size is very clearly stated on the nutrition information panel. Below are some examples of what a label could define as a serving size.

1 BABY FOOD JAR

1 SLICE OF GLUTEN FREE BREAD



1 CRISPBREAD

1 SNACK BAR

1 PACK OF VEGGIE CHIPS

1 ICE CREAM

Examples using the “per serving” column

EXAMPLE 1: BABY POUCH

NUTRITION INFORMATION		
Servings per package: 1	Average quantity	
Serving Size: 120g	Per serving	Per 100g
Energy	423 kJ	352 kJ
Protein	1.3 g	1.1 g
Fat, total	1.5 g	1.2 g
Carbohydrate	19.2 g	16 g
- sugars	11.7 g	9.7 g
Sodium	4 mg	3 mg

STEP 1: check the serving size

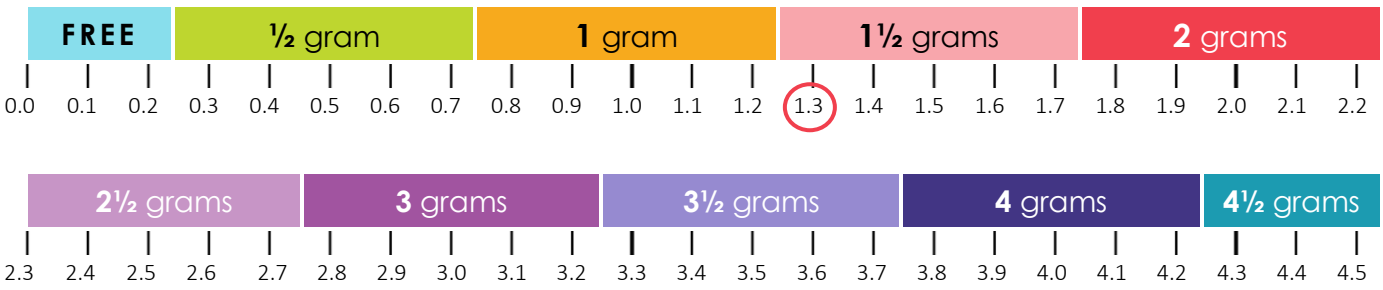
Find the nutrition information panel of the label. This example clearly states “Servings per package: 1”. A serving size is 120g which is the same as the net weight of the product, confirming that one pouch is equal to a serving. If you intend to use a full pouch the “per serving” column is the best option for calculating protein in this example.

STEP 2: find grams of protein “per serving”

Find protein on the left hand side and look across to the “per serving” column. In this example the product contains 1.3g protein “per serving” which is one pouch.

STEP 3: round to the nearest ½ gram of protein

To make it easier to add protein intake throughout the day it is recommended that protein is rounded to the nearest half. In this example, 1.3g should be rounded to the nearest half. If you find this calculation difficult you can use the scale below.



Answer: *one pouch is equal to 1½ grams (1.5g) of protein.*

Examples using the “per serving” column

EXAMPLE 2: SLICED BREAD

NUTRITION INFORMATION

Servings per Pack: 6 (10 SLICES AND 2 CRUSTS)

Serving Size: 67g (2 SLICES)

	AVERAGE QUANTITY PER SERVING	% DAILY INTAKE (PER SERVING)	AVERAGE QUANTITY PER 100g
ENERGY	690kJ	8%	1030kJ
PROTEIN, TOTAL	2.1g	4%	3.2g
- gluten	Not Detected		Not Detected
FAT, TOTAL	2.9g	4%	4.3g
- saturated	<1.0g	<4%	<1.0g
CARBOHYDRATE	31.0g	10%	46.2g
- sugars	2.2g	2%	3.3g
DIETARY FIBRE	2.3g	8%	3.4g
SODIUM	285mg	12%	425mg

STEP 1: check the serving size

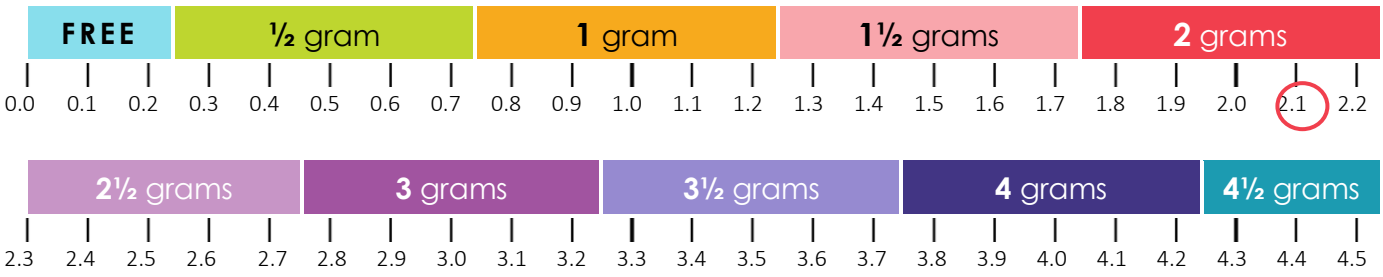
The label states that a serving size is 67g (2 slices).

STEP 2: find grams of protein “per serving”

Find protein on the left hand side and look across to the “per serving” column. A serving (2 slices) contains 2.1g protein.

STEP 3: round to the nearest ½ gram of protein

2.1g protein should now be rounded to the nearest half gram to make it easier to add up protein throughout the day.



Answer: *two slices of bread is equal to 2 grams (2.0g) of protein.*

Examples using the “per serving” column

EXAMPLE 3: SNACK BAR

Nutrition Information
(AVERAGE)
Servings per package: 6
Serving size: 22g (1 BAR†)

	quantity per serving	% daily intake ▲ per serving	quantity per 100g
ENERGY	380 kJ	4%	1730 kJ
PROTEIN	0.7 g	1%	3.4 g
FAT, TOTAL	1.9 g	3%	8.8 g
- SATURATED	0.7 g	3%	3.1 g
CARBOHYDRATE	17.4 g	6%	79.0 g
- SUGARS	7.1 g	8%	32.3 g
DIETARY FIBRE	0.1 g	0.3%	0.4 g
SODIUM	68 mg	3%	310 mg

†Bar weight is approximate and is only to be used as a guide. If you have any specific dietary requirements please weigh your serving.
▲ Percentage daily intakes are based on an average adult diet of 8700kJ.

STEP 1: check the serving size

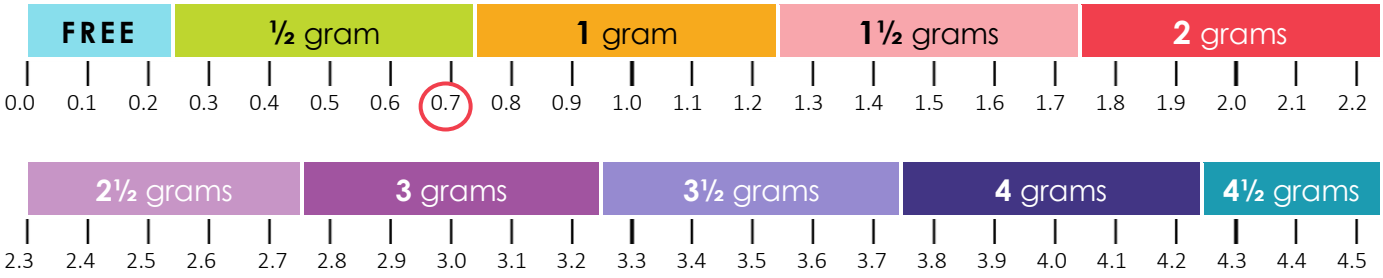
The label states that a serving size is 22g (1 bar).

STEP 2: find grams of protein “per serving”

Find protein on the left hand side and look across to the “per serving” column. A serving (1 bar) contains 0.7g protein.

STEP 3: round to the nearest ½ gram of protein

0.7g protein should now be rounded to the nearest half gram to make it easier to add up protein throughout the day.



Answer: *one bar is equal to ½ a gram (0.5g) of protein.*

Examples using the “per serving” column

EXAMPLE 4: BABY RICE CEREAL

NUTRITION INFORMATION

Servings Per Package: 13
Average Serving Size :105g
(Prepared as 15g of dry cereal + 90mL of water)

Average Quantity	Per Serving Prepared	Per 100g Prepared
Energy	244 kJ 58 Cal	232 kJ 55 Cal
Protein	1.1 g	1.0 g
Fat, total	0.2 g	0.2 g
Carbohydrates	13.1 g	12.5 g
-sugars	3.2 g	3.0 g
Sodium	6 mg	6 mg
Vitamin C	3 mg (10% RDI*)	3 mg
Iron	1.5mg (50% RDI*)	1.4 mg
Bifidus BL (Probiotic)	0.36 billion cfu	0.34 billion cfu

STEP 1: check the serving size

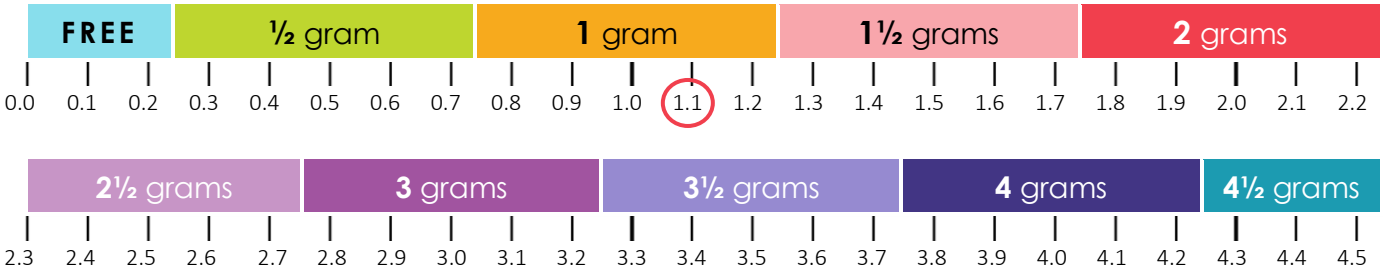
This “per serving” column is based on the prepared serving size. The “Average Serving Size” is 105g and based on the prepared recipe of 15g dry cereal + 90ml water. To ensure the serving size is correct 15g dry cereal or 105g prepared cereal (if made exactly as stated in the recipe) should be weighed. Dry cereal will give the most accurate results as it is not dependent on amount of water added.

STEP 2: find grams of protein “per serving”

Find protein on the left hand side and look across to the “per serving” column. In this example the product contains 1.1g protein “per serving” which is 15g dry powder or 105g prepared with water.

STEP 3: round to the nearest ½ gram of protein

For ease of calculations 1.1g should be rounded to the nearest half. See the scale below.



Answer: *15g dry cereal (or 105g cereal prepared exactly as stated in the recipe) is equal to 1 gram (1.0g) of protein.*

Using the “per 100g” column

The “per 100g” column can always be used to calculate protein however it will require weighing scales so may not always be practical if away from home.

It is necessary to use the per 100g column when:

- ☑ The serving size includes another ingredient that you are not planning to use
- ☑ The “per serving” column is not listed
- ☑ The “per serving” column is unclear or you are intending to eat more or less than the serving size.
- ☑ The “per serving” protein is listed as <1g however an exact amount of protein is listed in the per “100g column”

Here are some examples of when to use this column:

SERVING INCLUDES ANOTHER INGREDIENT

e.g. a serving of cereal is listed as prepared with skim milk however low protein milk will be used instead

SERVING SIZE DIFFERS TO PLANNED INTAKE

e.g. one large bag of chips contains 5.9 servings per pack however this is different to the size that will be eaten

NO PER SERVE COLUMN

e.g. only the “per 100g” column is listed and 100g (½ a pack) is more than will be consumed

SERVING SIZE <1g

e.g. a serving (one wafer) is listed as <1g but you need a more exact number. The “per 100g” column lists an actual number

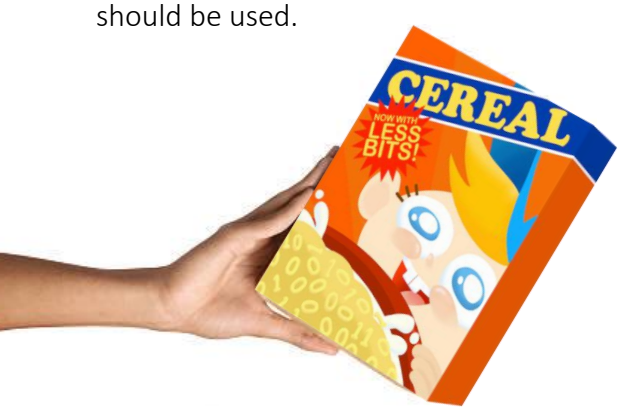
Using the “per 100g” column

There are two ways to use the “per 100g” column:

- ☑ To find out amount of food to weigh to provide one gram of protein
- ☑ To find out how much protein is in a specific amount of food

How much food should I weigh?

If you know how much protein you want to eat and need to know how much food to weigh this calculation should be used.



How much cereal should I weigh to give me one gram of protein?

100	÷	Protein “per 100g”	=	Amount equal to 1g protein

How much protein is in this?

If you have weighed out an amount of food and need to know how much protein is in it the calculation below should be used.

I’ve weighed out 30g cereal. How much protein is in this?

Protein “per 100g”	÷	100	X	Weight of food	=	Grams of protein

The examples on the next pages will help to explain this process.

Examples using the "per 100g" column

EXAMPLE 1: Cereal with skim milk

HOW MUCH TO WEIGH?

You would like to know how much to cereal to weigh to give one gram of protein without any milk.

STEP 1: find the grams of protein "per 100g"

Find protein on the left hand side and look across to the "per 100g" column to find that this product contains **7.4g** protein per 100g.

Nutrition Information
(AVERAGE)
servings per package - 10
average serving size - 35g (1 metric cup†)

	% daily intake ▲ per serving	per serve with 1/2 cup skim milk	quantity per 100g
ENERGY	6%	740 kJ	1550 kJ
PROTEIN	5%	7.2 g	7.4 g
FAT, TOTAL	0.1%	0.2 g	0.2 g
- SATURATED	0.1%	0.1 g	<0.1 g
CARBOHYDRATE	9%	34.7 g	80.7 g
- SUGARS	3%	9.3 g	8.1 g
DIETARY FIBRE	6%	1.8 g	5.1 g
SODIUM #	8%	249 mg	550 mg

Protein "per 100g"

STEP 2: use the calculation

Now that you know the grams of protein per 100g you can use your calculator to enter the formula below.

STEP 3: check the calculation

The answer on your calculator is the weight of cereal equal to 1 gram of protein. You should round to the nearest gram. If you find this difficult you can use the scale below to round up or down based on the number after the dot. This number rounds to 14 grams (14g).

STEP 4: weigh the product

If you want to eat 1 gram of protein simply weigh out 14g cereal. If you want to eat more than one gram you can multiply the number on your calculator by how many grams of protein you are planning to eat. For example:

- **2 grams**: multiply by 2 i.e. 2 x 14g = 28g
- **3 grams:** multiply by 3 i.e. 3 x 14g = 42g
- **4 grams:** multiply by 4 i.e. 4 x 14g = 56g

Examples using the "per 100g" column

Example 2: Chips

HOW MUCH TO WEIGH?

The serving size is 20g (about 13 chips) however you would like to eat one gram of protein.

STEP 1: find the grams of protein "per 100g"

Find protein on the left hand side and look across to the "per 100g" column to find that this product contains **1.4g** protein per 100g.

STEP 2: use the calculation

Now that you know the grams of protein per 100g you can use your calculator to enter the formula below.

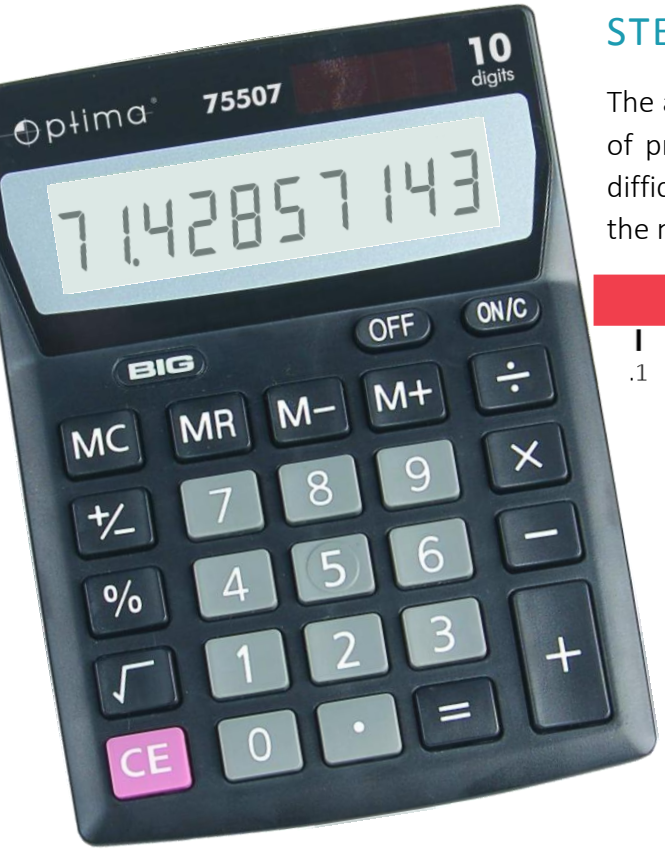
NUTRITIONAL INFORMATION

SERVINGS PER PACKAGE: 5.9

SERVING SIZE: 20g (About 13 Chips)

	Average Quantity per Serving	% Daily Intake (per Serving)	Average Quantity per 100g
ENERGY	378 kJ	7 %	1890 kJ
PROTEIN, TOTAL	0.3 g	4 %	1.4 g
FAT			
-TOTAL	3.2 g	9 %	15.9 g
-SATURATED	0.3 g	4 %	1.6 g
-TRANS	<0.1 g		0.1 g
-POLYUNSATURATED	0.4 g		2.0 g
-MONOUNSATURATED	2.4 g		12.2 g
CARBOHYDRATE	13.6 g	5 %	68 g
-SUGARS	2.2 g	2 %	11.1 g
DIETARY FIBRE	1.0 g	8 %	5.4 mg
SODIUM	134 mg		670 mg

Protein "per 100g"



STEP 3: check the calculation

The answer on your calculator is the weight of chips equal to 1 gram of protein. You should round to the nearest gram. If you find this difficult you can use th e scale below to round up or down based on the number after the dot. This number rounds to 76 grams (76g).

STEP 4: weigh the product

If you want to eat 1 gram of protein simply weigh out 76g of chips. If you want to eat more than one gram you can multiply the number on your calculator by how many grams of protein you are planning to eat. For example:

- **½ gram:** multiply by 0.5 i.e. 0.5 x 76g = 38g
- **1½ grams:** multiply by 1.5 i.e. 1.5 x 76g = 114g

Examples using the “per 100g” column

Example 3: Rice Cakes

HOW MUCH PROTEIN IN THIS?

You would like to know how much protein is in one rice cake however this label only has a “per 100g” column.

STEP 1: weigh a rice cake

You find the weight of a rice cake to be **13g**.

STEP 2: find the grams of protein “per 100g”

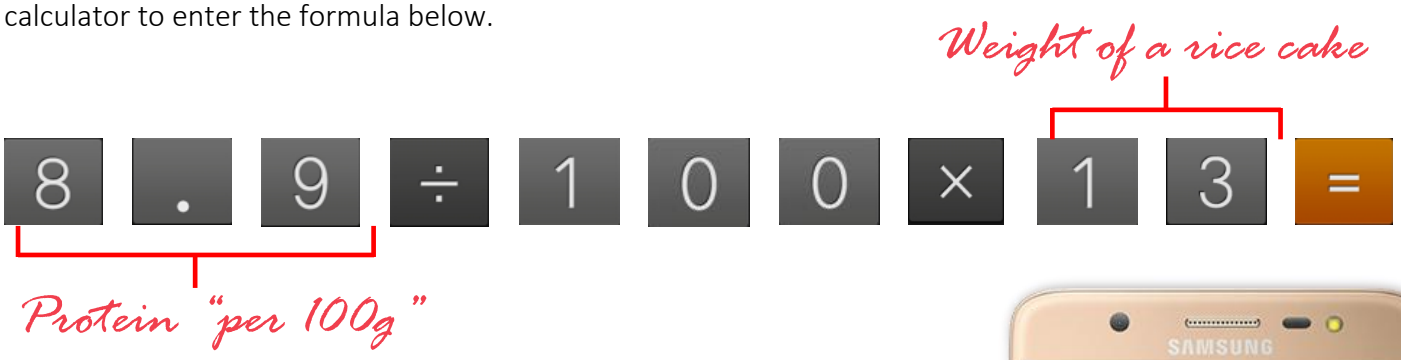
This product contains **8.9g** protein “per 100g”.

Nutrition Information

	Average Quantity per 100g
Energy	1690kJ (404 Cal)
Protein, total	8.9g
- gluten	0mg
Fat, total	8.3g
- saturated	1.5g
Carbohydrate	70.5g
- sugars	4.2g
Sodium	808mg

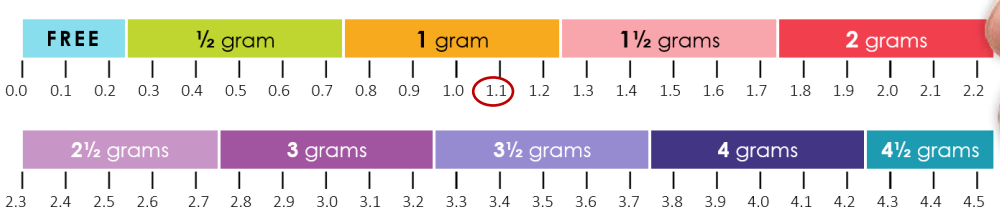
STEP 3: use the calculation

Now that you know the weight of a rice cake and the grams of protein “per 100g” you can use your calculator to enter the formula below.



STEP 4: check the calculation

The answer on your calculator is the amount of protein in one rice cake. You should round to the nearest ½ gram. If you find rounding difficult you can use the scale below to round the number after the dot down or up.



In this example one rice cake contains one gram (1g) of protein.

Examples using the “per 100g” column

Example 4: Wafers

HOW MUCH PROTEIN IN THIS?

You want to know how much protein in one wafer. The serving size is listed as 22g however it is unclear if this is the weight of a wafer. Also a serve is <1g but you would like a precise number. The nutrition information panel shows grams of protein in the “per 100g” column.

NUTRITION INFORMATION Servings Per Pack: 6 Serving Size: 22g		
	Ave. Quantity Per Serving	**Ave. Quantity Per 100g**
Energy	480kJ (115kcal)	2180kJ (522kcal)
Protein	<1g	3.4g
Fat, total	6.2g	28g
- saturated	3.7g	17g
Carbohydrate	14g	63.5g
- sugars	10g	46.5g
Sodium	18m	84mg

STEP 1: weigh a wafer

You find the weight of a wafer to be **22g** (the serving size).

STEP 2: find the grams of protein “per 100g”

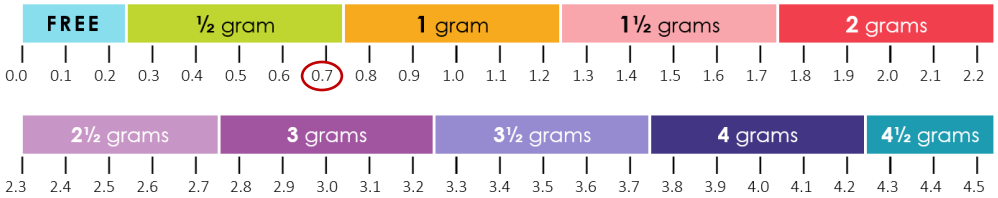
This product contains **3.4g** protein “per 100g”.

STEP 2: Use the calculation

Now that you know the weight of a wafer and the grams of protein “per 100g” you can use your calculator to enter the formula below.

STEP 3: check the calculation

The answer on your calculator is the amount of protein in one wafer. You should round to the nearest ½ gram. If you find rounding difficult you can use the scale below to round the number after the dot down or up.



In this example one wafer contains ½ gram (0.5g) of protein.

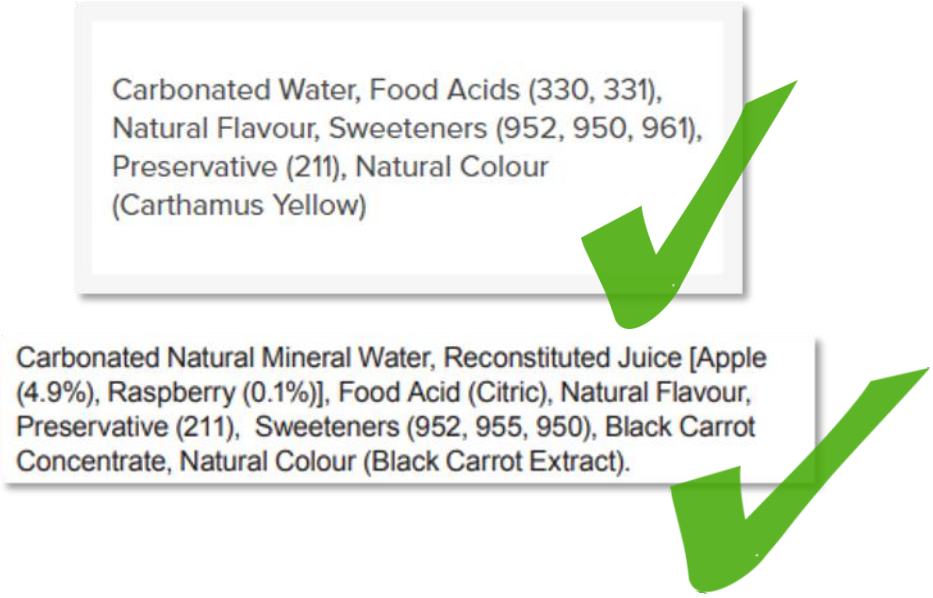
Artificial Sweeteners

Some artificial sweeteners that are added to foods contain phenylalanine (Phe) and should be avoided. These sweeteners are mainly used in diet drinks, sugar free lollies, chewing gum and some medications. It is important to check the ingredient lists of diet and sugar free foods to ensure that the following Phe containing artificial sweeteners are not added.

- Aspartame (additive 951)
- Acesulphame-Aspartame (additive 962)
- Nutrasweet
- Equal
- Canderal

Below are some examples of products that should be avoided.

Not all diet or sugar free soft drinks need to be avoided. If the product does not list sweeteners 951, 962, Nutrasweet, Equal or Canderal the product is okay to have.



Question 1

Use the nutrition information panel to the right to answer the questions below.

a) What is a "serving" of this product?

b) How many grams of protein are in a serving?

c) Can you round this to the nearest half gram?

NUTRITIONAL INFORMATION

Servings per package: 5
Serving size: 22 g (1 bar)

	Avg. Quantity per Serving	Avg. Quantity per 100 g
Energy	380 kJ (91 Cal)	1730 kJ (413 Cal)
Protein	0.7 g	3.1 g
- gluten	0 g	0 g
Fat, total	2.0 g	9.1 g
- saturated	0.8 g	3.5 g
Carbohydrate	16.8 g	76.2 g
- sugars	4.8 g	22.0 g
Dietary fibre	1.1 g	5.1 g
Sodium	21 mg	96 mg
Potassium	24 mg	109 mg

Question 2

Nutrition Information
(AVERAGE)

servings per package - 10
average serving size - 35g (1 metric cup†)

	% daily intake ▲ per serving	per serve with 1/2 cup skim milk	quantity per 100g
ENERGY	**6%**	740 kJ	1550 kJ
PROTEIN	**5%**	7.2 g	7.4 g
FAT, TOTAL	**0.1%**	0.2 g	0.2 g
- SATURATED	**0.1%**	0.1 g	<0.1 g
CARBOHYDRATE	**9%**	34.7 g	80.7 g
- SUGARS	**3%**	9.3 g	8.1 g
DIETARY FIBRE	**6%**	1.8 g	5.1 g
SODIUM #	**8%**	249 mg	550 mg

Use the nutrition information panel to the left to answer the questions below.

a) How many grams of this product (without milk) is equal to 1g protein

b) How much should you weigh if you wanted to eat 2g protein?

Question 3

NUTRITIONAL INFORMATION			
SERVINGS PER PACKAGE: 5.9 SERVING SIZE: 20g (About 13 Chips)			
	Average Quantity per Serving	% Daily Intake (per Serving)	Average Quantity per 100g
ENERGY	378 kJ	7 %	1890 kJ
PROTEIN, TOTAL	0.3 g	4 %	1.4 g
FAT			
-TOTAL	3.2 g	9 %	15.9 g
-SATURATED	0.3 g	4 %	1.6 g
-TRANS	<0.1 g		0.1 g
-POLYUNSATURATED	0.4 g		2.0 g
-MONOUNSATURATED	2.4 g		12.2 g
CARBOHYDRATE	13.6 g	5 %	68 g
-SUGARS	2.2 g	2 %	11.1 g
DIETARY FIBRE	1.0 g	8 %	5.4 mg
SODIUM	134 mg		670 mg

Use the nutrition information panel above to calculate how many grams of protein are in 37g of chips

Question 4

Nutrition Information		
SERVINGS PER PACKAGE:		25
SERVING SIZE:		45g
(prepared as 5g of dry cereal and 40mL of water)		
	Avg. Quantity per Serving prepared cereal	Avg. Quantity per 100g prepared cereal
ENERGY	75kJ	170kJ
PROTEIN	0.3g	0.8g
FAT, TOTAL	0.2g	0.4g
CARBOHYTRATE	3.8g	8.5g
- SUGARS	0g	0g
DIETARY FIBRE	0g	0g
SODIUM	1mg	2mg
IRON	1.1mg	2.5mg

Time for a hard question…. If you are able to answer all parts of this question correctly you have mastered label reading!

a) How many grams of dry cereal are in a serving?

b) How many grams of protein are in a serving?

c) How many grams of protein in 10g dry cereal?

d) How many grams of prepared cereal is equal to 1g protein?

e) How many grams of protein are in 80g prepared cereal?

*ANSWERS - **Question 1:** a) 22g (1 bar), b) 0.7g protein, c) 0.5g protein (½ gram), **Question 2:** a) 14g (round up from 13.5g), b) 28g, **Question 3:** 0.5g protein, **Question 4:** a) 5g, b) 0.5g (round up from 0.3g), c) 0.5g (round down from 0.6g), d) 125g, e) 0.5g (0.64g rounded down).*