The PKU Handbook

ASIEM gratefully acknowledges the financial support of SHS, Vitaflo and Abbott, which made the production of this handbook possible.

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Acknowledgements

This handbook has been prepared by the Dietitians Working Party of the Australasian Society for Inborn Errors of Metabolism (ASIEM), a special interest group of the Human Genetic Society of Australasia (HGSA).

Thanks to the people with PKU and families of people with PKU who contributed significantly to the handbook evaluation, focus group discussions and feedback on the first draft. They made sure useful information was included. Thanks too to those who organised and ran the focus groups.

We would especially like to acknowledge the contribution of editor, Barbara Dennison.

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Printed by Snap Printing, Eastwood

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Chapter 1

What is PKU?

Finding out about PKU

PKU, or phenylketonuria, is a treatable condition. People with PKU cannot break down foods containing protein in the usual way. When PKU is detected early and treated with a special diet, children are able to reach their full potential.

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

This handbook explains the important things about PKU and how it can be managed, and gives details of the support that's available. It also includes information, advice and encouragement from people who live with PKU.

Managing PKU is not about getting sick children better, it's about keeping healthy children healthy. When you get used to PKU and managing the diet, it isn't as overwhelming or daunting, and you can get on with raising your child.

What causes PKU?

PKU is an inherited disorder, caused by a faulty gene. Around one in 15,000 babies born in Australia and 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. The glossary on page 147 gives simple explanations of all medical terms used in this handbook.)

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)**, 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 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 phenylalanine in the usual way, it builds up in the blood and damages the brain. People with PKU need phenylalanine, but only a *small amount*. (Phenylalanine intake is explained in more detail in chapter 6.)

What is PKU?

How is PKU detected?

All babies born in Australia and New Zealand are tested for PKU soon after birth, usually the second or third day. A blood sample is taken from a needle prick on the heel, and the phenylalanine 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 given a special formula (also called a supplement) to lower the phenylalanine to a safe level. Before birth, the mother's blood circulation can deal with the baby's phenylalanine and the level stays within the normal range. Immediately after birth it starts to rise.

The period of time between birth and finding out your baby has PKU is too short to cause damage. Children with PKU treated from early infancy are able to reach their full potential.

Growing up with PKU

Maintaining the diet in childhood is essential to prevent damage to the growing brain. It is recommended that the diet is 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 PKU clinic doctor if you're uncertain.

Without treatment, babies with PKU would show signs of slow development by the end of their first year. If left untreated, PKU leads to severe brain damage.

Will other children in the family 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.

Chapter 1

What is PKU?

What is the treatment for PKU?

PKU is treated with a low protein diet and a special nutritional supplement, which need careful monitoring. Regular blood tests to measure the phenylalanine level and regular attendance at the PKU clinic are part of the treatment.

For healthy growth we all need to eat carbohydrate, fat, protein, minerals and vitamins. People with PKU can't break down the large amount of phenylalanine in protein foods, so the amount of protein in the diet is *restricted*. A special supplement replaces the protein, minerals and vitamins that would normally be in the diet. The supplement contains all the amino acids except phenylalanine. People with PKU obtain the other nutrients they need from their food intake. See the Five Food Groups at the end of this chapter, which shows how the PKU diet varies from a normal diet.

'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 PKU team was wonderful.'

Feeding your newborn baby

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

The PKU clinic team – usually a dietitian, doctor, nurse, laboratory scientist and perhaps a social worker – 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 certain baby foods. Later on, they can eat a wider range of low protein foods, including special bread and pasta, and other special low protein products. The PKU clinic dietitian will help you manage the diet.

The amount of protein and phenylalanine intake from food and supplement is worked out by measuring the blood level and altering the intake accordingly.

What is PKU?

Safe phenylalanine levels

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

What is the right phenylalanine level?

PKU clinics differ on what they consider to be 'acceptable' levels of phenylalanine. From time to time phenylalanine levels do vary. 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 eight years old Phe levels should be 100–350 micromoles per litre (µmol/L)
- for older children, teenagers and adults, most clinics recommend a maximum level of 450 µmol/L – however, some clinics accept levels up to 600 or 700 µmol/L (while this is not ideal), and other clinics say people with PKU should aim to keep their levels around 100–350 µmol/L for life
- women planning a pregnancy or who are pregnant need to have lower levels to protect the baby (see PKU and Pregnancy, chapter 12)
- be guided by your clinic.

How is PKU inherited?

Genes

PKU is caused by a faulty gene. Genes carry hereditary information about body processes and traits, such as blood group and hair colour, from parents to their children. Children inherit one set of genes from each parent. Arranged in pairs, these genes are replicated in every cell of the child, and carry the 'blueprint' for each function and characteristic of their body.

When genes are faulty

A person who has a faulty gene, but is not affected by it, is called a 'carrier'. Everyone carries some faulty genes.

In PKU, as for most other enzyme deficiencies, as long as only one of a pair of genes is faulty, there are no noticeable effects. But if both genes are faulty, problems will arise. When two people, each with the same faulty gene have children, their child may be affected.

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What is PKU?

PKU: a genetic disorder

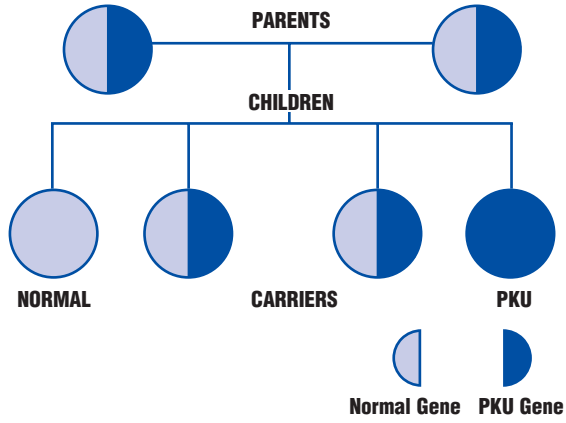
Children born with PKU have inherited a faulty PKU gene from each parent. Around one in 50 or 60 of us has a faulty PKU gene among the thousands of genes each of us possesses, yet only one in 15,000 Australians or New Zealanders has inherited two defective genes and has PKU. Boys and girls have an equal chance of inheriting PKU.

The faulty gene in PKU is one that controls the enzyme, phenylalanine hydroxylase (PAH), needed to break down phenylalanine in the body. With two faulty PKU genes in their cells and no regular ones, people with PKU cannot make functioning phenylalanine hydroxylase in sufficient amounts. This affects the way the body deals with phenylalanine.

What does this 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 faulty gene, each egg or sperm cell will carry *either* a faulty or a regular gene. If, by chance, both parents carry one copy of the same faulty gene, any baby conceived has:

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



If both parents carry one faulty PKU gene there is a 1 in 4 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 PKU is very low – from 1 in 100 to 1 in 120.

Testing for possible carrier status in a family with no history of PKU is difficult and not always accurate. 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.

Why is the condition called phenylketonuria?

The lack of PAH means phenylalanine cannot be properly converted for use in the body, so blood levels of phenylalanine rise, and eventually the phenylalanine 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, phenylketones are excreted, so the condition is called Phenylketonuria.

PKU: the science behind the condition

The phenylalanine that builds up in PKU comes from protein. All protein is composed of chemicals called amino acids. One of these is *phenylalanine or Phe*.

Amino acids in protein are joined to one another, much like beads on a string. Every so often, one of the beads is phenylalanine.

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

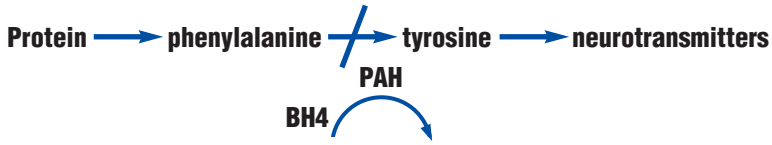
What is PKU?

individual amino acids are absorbed into the blood stream, and processed for use in building muscle, making other chemicals in the body, or to provide energy.

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

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

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



Tetrahydrobiopterin (BH4)

BH4 is important too. Babies found by newborn screening to have a high blood phenylalanine level may be admitted to hospital for a test to find out for sure where the problem is, whether it is with *phenylalanine hydroxylase (PAH)* or *BH4*.

Some people with PKU, (and a problem therefore with PAH) may be helped by treatment with BH4. At the moment, the expense of this treatment prohibits its widespread use, but it is hoped that this problem will be overcome in the near future.

Tyrosine

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

Tyrosine is generally obtained from food, so people with PKU who are *not* on treatment lack tyrosine. Tyrosine is included in the Phe-free formula or supplement given to people with PKU. Sometimes it is given separately.

FIVE FOOD GROUPS

Five food groups

- fruit and vegetables
- breads and cereals
- meat, chicken, fish
- milk and diary

- fats and sugars

PKU food guide

- fruit and vegetables
- low protein bread, flour and pasta

- protein supplement
- fats and sugars

Chapter 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
- can I have another child?
- treating PKU: the role of the family

Chapter 2

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

Your feelings

Accepting the diagnosis of PKU can be difficult because your baby looks well, even perfect. 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 it may not be clear at first what PKU really means.

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

As the good news that PKU can be controlled starts to balance some of the initial distress, the amount of information about the special 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, think about the things that will be different and what your child will miss out on. Most parents ask the question, why me or 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 make-up finding each other. It can help to remind yourself that the PKU is not something you could have avoided, nor is it the result of 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 phenylalanine 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 worry or have doubts about how you will cope.

There's plenty of time to learn about the diet before your baby starts solids, and you can take one step at a time. It often takes the first year or more to gain confidence in managing your child's PKU.

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

Chapter 2

Sharing the experience

One thing that helps during this time is regular contact with the PKU clinic team through visits and phone calls, if needed. 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 where possible. 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.

A team approach

PKU is not something parents can manage on their own. Even though it can sometimes be difficult to accept guidance on something that seems basic, such as feeding your baby, following the instructions of the PKU team is crucial.

A child's intellectual development is put at risk if the family is unable or unwilling to follow the advice of the medical team and their PKU is not well managed. 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

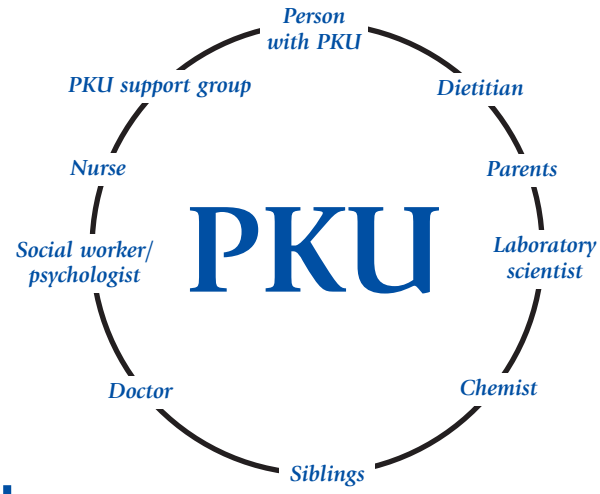
One of the challenges of having a child with PKU is that, because it is rare, few people have heard of it. Finding ways of explaining PKU as simply as possible to yourself, your family, your child and interested others will evolve over time and as the need arises.

Some people will feel ready to tell family and friends about the diagnosis soon after the birth, but you may want to avoid telling too many people until you have adjusted to the diagnosis and the extra responsibility it brings. 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. (Chapters 9 and 18 look at giving specific information to carers, preschool and school teachers, 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 alright, it's not that bad, the more I was reassuring myself it was OK.'

Chapter 2

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



The PKU support circle: you will not be alone in managing your child's PKU

'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.'

Telling the grandparents can be difficult, but it is important to do this early if you can, as ideally, the whole family needs to pull together.

The most important person you will need to talk about PKU with will be your child (see chapter 8).

Can I have another child?

Parents are often concerned about whether they can have more children. There's a one in four chance that another child born to the same parents will also have PKU. (Chapter one explains how PKU is inherited.) This is an individual decision, however the management of PKU is so successful that many parents feel there is no reason not to have another child.

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

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 clinic team will help you at each stage and in any way they can.

Successful treatment of PKU includes the following:

1. A diet low in phenylalanine
The amount of protein allowed in your child's diet is calculated with you regularly. This is worked out by looking at their phenylalanine levels, age and growth.

2. A daily dietary supplement
This is also calculated specifically for your child's needs. It may be given in the form of a formula, drink or gel. The supplement should be taken several times throughout the day to maximise its effect.

3. Regular monitoring of blood samples
Blood levels are the most accurate way to measure phenylalanine 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 information and support. Visits are likely to vary from once a week to six weekly. As your understanding and confidence grows, the time between clinic visits will increase. Most school age children (with stable results) visit the clinic every three to six months. Attending a PKU clinic is also a requirement for receiving financial assistance from the government (in Australia).

5. Attending education sessions
Individual clinics may provide ongoing educational sessions for managing PKU at specific times or ages. These are more frequent in the early years when there are more changes in diet. Attending these sessions helps increase your understanding of the condition and gives you the opportunity to establish networks with other families and gain from the experience of others.

6. Keeping supplies up-to-date
Part of your responsibility is to make sure you have an up-to-date supply of prescriptions, newborn screening cards and blood taking devices.

Chapter 2

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

By now you have probably met the different members of your PKU team. Each has a specific role in helping you meet your child's needs. Your clinic will provide contact details for the following:

PKU consultant _____

Metabolic doctor on call _____

Metabolic/PKU dietitians _____

Metabolic/PKU nurse _____

Clinic bookings _____

Newborn screening _____

Chapter 3

Feeding your baby

- breastfeeding
- formula feeding
- Phe-free formulas
- how many feeds does my baby need?
- how to express breast milk
- where to go for help
- looking after yourself
- what to do if you are unwell
- stopping breastfeeding
- how to clean and sterilise feeding equipment

Chapter 3

Feeding your baby

Breastfeeding

Breast milk is 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 with PKU and keep the blood Phe at a safe level. Breast milk contains much less Phe than infant formula. However, breast milk alone contains too much Phe for babies with PKU. A Phe-free formula is given by bottle to take the edge off your baby's appetite before breastfeeding.

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

For the first few days your baby may be given just the Phe-free formula and no breastfeed or standard infant formula. This will allow the high phenylalanine blood level to come down more quickly.

The clinic 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 your supply of breast milk. (See How to Express Breast Milk later in this chapter.)

Starting breastfeeds

After the first few days, breastfeeds are given **after** a measured amount of Phe-free formula. Most babies with PKU need about half the usual amount of breast milk, but this varies. If your baby is hungry offer more breastfeeds after their Phe-free formula.

Starting formula feeds

After the first few days, standard infant formula is usually given *before* the Phe-free formula. If your baby is hungry, offer more Phe-free formula. Your clinic will advise you how to do this.
Most babies quickly learn to cope with the combination of bottle and

The PKU team will work out 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. Usually the breast is offered after the Phe-free formula has been given. Formula-fed babies are usually given the Phe-free formula last.

breastfeeding or the two different formulas. 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 safe 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.

Phe-free formulas

Phe-free formulas contain all the nutrients needed for growth, except phenylalanine. Your PKU clinic will prescribe the appropriate formula for your baby.

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

To prepare the formula:
- 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
- unless your clinic team advises you to do so, it is not necessary to add anything to your baby's formula.

To store the formula:
- make up each bottle as required or you can make up 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 feed after 24 hours.

At feed time

1. Carefully measure the amount of formula required into a feeding bottle.
2. Warm the bottle in a jug of hot water (not boiling). It is best not to heat the bottle in the microwave as uneven heating can cause hot patches which may burn your baby's mouth.
3. 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 not feel hot.
4. If your baby doesn't drink all of the formula, record what has been drunk and throw out what is left after an hour.

Feeding your baby

What to do when you're going out

Take the refrigerated feeds in an insulated bag or polystyrene container with an ice pack to keep them cool. Otherwise, take boiled water and formula with you to make up the feed when you need it. A wide-necked vacuum flask of hot water can be used to warm the feeds.

How many feeds does my 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
- a new feed starts when an hour or more has passed since the last feed finished
- your baby may have extra breastfeeds in addition to the Phe-free formula and usual breastfeeds
- provided your baby is well, nothing apart from Phe-free formula and breastfeeds or infant formula and Phe-free formula is needed until around six months of age – any extras, even water, can affect breastfeeding
- babies should not be given herbal or medicinal teas, and extra vitamins and minerals are not needed.

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 PKU clinic team.

Talk to the PKU 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 from your breasts to keep up your supply.

You can express your milk by hand or with a breast pump – using an electric or hand pump. Most pharmacies and some hospitals sell hand pumps and hire electric pumps.
You'll need to express six to eight times a day, expressing every three to four hours during the day and less at night.

1. First, wash your hands.
2. If you're expressing by hand, gently squeeze the ducts behind the areola (dark area around your nipple) in a steady rhythm. Collect the milk in a sterile cup then transfer it to a sterile bottle.
3. If you're using a pump make sure the pump parts that come into contact with the milk are sterile.
4. Your milk can be stored in bottles and frozen for later use. It will keep for four months in a deep freeze, or two weeks in the freezer section of a refrigerator.
5. Milk from several expressions can be added to the same bottle. Use a new bottle at least every 24 hours or when the bottle is three-quarters full.
6. Label the bottle 'expressed milk', with the date and time it was expressed.

Where to go for help

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

- the maternal child health nurse (Australia), or Plunket Nurse (New Zealand) or breastfeeding advisor at your hospital
- your health centre nurse
- PKU clinic nurse
- a midwife
- a lactation counsellor
- the Australian Breastfeeding Association or La Leche in New Zealand (web address and phone numbers can be found in PKU Resources, chapter 17).

Explain that your baby has PKU so they will understand why you need advice about expressing milk rather than 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 milk supply
- try to eat healthy snacks if you can't manage full meals
- limit your tea, coffee and cola intake to less than four cups a day
- 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 little you may be producing – your supply will adjust to your baby's feeding demands
- babies with PKU only need about half their feed as breast milk, so some drop in supply is no cause for concern.

Feeding your baby

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 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 so you don't lose 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 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. Paracetamol (Panadol) may help relieve pain. Antibiotics may also be needed.

What to do if you are unable to breastfeed

Seek advice from your PKU 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, or
- 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, make it up according to the directions and give 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
- keep in close contact by daily phone calls with your PKU clinic team until you return to your usual routine.

Stopping breastfeeding

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

If you wish to stop breastfeeding, it is best to plan it with the PKU clinic team and do it over two to three weeks.

How to clean and sterilise feeding equipment

Cleaning

Make sure bottles, teats and caps are thoroughly clean before sterilising. Wash them in hot soapy water and rinse well. Use a brush inside the bottles and rub the teats between the palms of your hands to help remove the milk residue. Squeeze water through the feeding holes.

Sterilising

This can be done in one of three ways: boiling, steaming, or using a commercial sterilising solution. Remember to wash your hands thoroughly before handling sterilising equipment.

Boiling method

1. Place the bottles and caps in a large saucepan. Leave the teats out at this stage.
2. Cover with water, making sure the bottles are full of water and completely covered. Cover the saucepan with a lid.
3. Bring to the boil. After two minutes, add the teats and continue boiling for a further three minutes.
4. Turn off the heat and allow to cool.
5. Bottles and teats may be left in the covered saucepan until the next feed, or assembled and stored in a clean, dry place.
6. Before removing the sterilised items, wash and dry your hands. Drain the water off each item and remove from the saucepan using tongs. Avoid touching the tops of the bottles and the tips of the teats with your hands.
7. To store sterilised bottles, invert the teat into the bottle and seal with the cap and disc. Store in the refrigerator or on a clean tray, covered with a clean cloth.

Steaming method

Easy-to-use electric sterilisers or microwave units designed specifically for this purpose are available from pharmacies or can be purchased on the web.

1. Place the feeding equipment into the microwave unit or electric steriliser.
2. Add the amount of water specified in the instructions.
3. Cover the unit.
4. Follow the manufacturer's instructions to make sure all items are sterilised effectively.

Feeding your baby

Using sterilising solutions

Several sterilising solutions are available for sterilising feeding bottles and teats. As well as the solution, you will need a sterilising unit or a large bucket and lid.

1. Follow the manufacturer's instructions when making up the solution, making sure the proportions of water and sterilising solution are accurate.
2. After cleaning, place the bottles, caps and teats into the diluted sterilising solution, making sure they are completely covered.
3. Leave the feeding equipment in the solution for the time recommended by the manufacturer.
4. Bottles, teats and caps can be stored in the sterilising solution until required. If you add any extra items, re-time the whole sterilising process.
5. Before removing the sterilised items, wash and dry your hands thoroughly. Avoid touching the tops of the teats or bottles. Drain well and use them as soon as possible. Do not rinse in water first.
6. Remake the liquid in the sterilising bath every 24 hours.
7. Keep the solution away from children.

Chapter 4

The PKU diet: understanding the protein supplement

- different kinds of supplements
- taking the supplement
- when your child refuses the supplement
- taking the supplement when you're away from home

Chapter 4

The PKU diet: understanding the protein supplement

THE THREE PARTS OF THE PKU DIET

1. **Formula or supplement**
2. **Counted Phe from food**
3. **Phe-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 they don't get from food. For people with PKU, the supplement is vital. It provides all the essential amino acids (except phenylalanine), tyrosine, vitamins, minerals and trace elements.

Different kinds of supplements

A variety of supplements is 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 lifestyles and preferences. Your dietitian will advise which are suitable for you or your child.

Babies are fed Phe-free formula; toddlers, older children and adults have more supplement options:

- **powder** comes in cans or sachets and is mixed into a drink with water, with or without flavouring – in some cases extra vitamin and mineral supplements need to be taken as well
- **gel or thick paste** can be eaten with a spoon or taken as a low volume drink – flavoured or unflavoured products are available
- **liquid** – this comes as a single serve, ready to drink
- **capsules or tablets** – extra vitamin and mineral supplements are needed with these
- **bars** – vitamin and mineral supplements are needed.

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

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

The PKU clinic doctor will provide a prescription for the supplement, which is available from pharmacies. As PKU is a rare condition and the supplements are expensive, your local pharmacist will not have them in stock. Some may not have heard of the product and may initially say they cannot get it. The pharmacist may need to make direct contact with the company that produces the product.

The PKU diet: understanding the protein supplement

Chapter 4

Supplement supply

Choosing a reliable pharmacy, conveniently located close to home or work is important. Go to the same pharmacy each time, and remember to place your order before you run out. It can take anywhere from a few days to a few weeks for a pharmacy to obtain your supplement.

You will have to pay the standard prescription charge for each prescription. With government subsidies, the supplements cost you only a small amount, but they are extremely expensive to produce, so try to minimise waste.

Taking the supplement

'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.'

It is best to take the supplement with meals in several doses spread throughout the day – ideally over 12 hours. Also:

- follow the supplement prescription carefully
- if you weigh your supplement, check that your scales are accurate – do this by measuring 50 ml of water and weighing it, it should weigh 50 g
- if you use tablespoons to measure your supplement, make sure you're using a standard metric measure – 1 tablespoon = 20 ml in Australia, 1 tablespoon = 15 ml in New Zealand
- if your prescription is for scoops, use the one provided in the can of supplement or formula
- measure level rather than rounded scoops or tablespoons.

Tips for parents of young children

The full amount prescribed should be taken every day. This is important for growth and development, and blood Phe levels. 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 foods you are counting in your child's diet – offer small amounts throughout the meal if your child won't drink it at the beginning
- 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 13 – Dental Care and PKU)
- is best given from a sipper cup or cup rather than an infant formula bottle from when your child is about twelve months old.

Chapter 4

The PKU diet: understanding the protein supplement

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 **covered cup with a straw** to reduce the smell
- **flavoured** with cordials, lemonade, fruit flavoured topping, coffee creamers, vanilla, peppermint essence, and tomato juice.

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

Make sure all the supplement is drunk, including the sediment at the bottom, which contains the important minerals, vitamins and tyrosine. Sometimes drinking the supplement is difficult for children, and these times are a challenge for parents.

When your child refuses the supplement

'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.'

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
- there is not enough importance attached to having the supplement
- someone may be making negative comments about the supplement
- the supplement is mixed with too much liquid so the amount is too much for your child to manage – or maybe there's too little liquid, making 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 different cups can make the supplement more acceptable. Sometimes changing to an alternative supplement is helpful.

Taking the supplement when you're away from home

With young children, when you're eating out, take the supplement with you for your child to drink with the meal. If this is not practical, give some before and after the meal.

The PKU diet: understanding the protein supplement

Use insulated carry bags to keep drinks cool. Small thermos containers are another way to keep the supplement cool.

Some people take extra supplement with them when they're away from home in case of an unexpected delay, such as car trouble.

At school and work

Encourage your child to take some protein supplement to school to drink throughout the day. Drink bottles with straws are popular, as are sport bottles. To keep the drink icy cold on hot days, freeze some water overnight, then top up the container with supplement in the morning and the drink will be ready at lunchtime.

If taking the supplement at school is too difficult, give some before and after school and at bedtime. (see Chapter 13 – Dental Care and PKU)

If you prefer your supplement chilled, and there's no fridge available where you work, make it the night before and take the lunchtime supply in a thermos to keep it cool.

Travelling

You'll find more information about how to manage the diet and supplement when you're away from home in the quiz at the end of this handbook. If you're travelling overseas, you'll need a letter for customs explaining what the supplement is and why it is required. You'll find a sample letter in chapter 15.

'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.'

Chapter 5

Blood samples

- collecting blood samples
- how often are blood samples needed?
- how to take a blood sample
- trouble shooting
- tips for success
- involving children in their blood tests
- where are blood samples sent (NZ and Australia)?
- what affects phenylalanine levels?

Chapter 5

Blood samples

Collecting blood samples

Phenylalanine (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. Nursing staff will take the samples until you feel you're ready to learn how to do it yourself. 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.

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

Blood testing is the only accurate way to measure phenylalanine levels in the blood. Tests are taken regularly – your PKU 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 your baby's diet becomes established, blood tests may be needed twice weekly or weekly
- once the diet is established, twice weekly, weekly, or fortnightly tests are generally taken for the first 12 months – this is a period where your baby will experience significant growth and change of diet
- after the first 12 months, weekly, fortnightly or monthly tests continue throughout life – taking the blood tests on the same day of the week, and, if monthly, putting a date on the calendar each month, e.g. your child's birthday, or yours, helps remind you
- additional blood samples may be needed after (or during) an illness or when the levels are too high or too low.

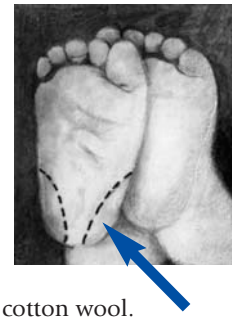
How to take a blood sample (from the heel or finger)

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 is 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 is important that the blood completely soaks through the card and that blood drops are not placed on top of each other.

Blood samples

1. Fill in the details on the filter paper (newborn screening card) using a ball point 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')
- filter paper (newborn screening card)
- tissues or cotton ball
- Band-Aid

3. Wash your hands and make sure the heel or finger has been washed and dried.
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 to take the blood sample from – (see diagrams).
6. Press 'pen' or other device firmly against the heel or finger and push down till 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. 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 in the air for *at least four hours*.
13. When it is *fully dry*, wrap the card in plain paper.
14. Place in an envelope and send to your Newborn Screening Centre at the address on the card (or to the address on the label supplied).

Trouble shooting

Common problems with samples, which generally mean the laboratory is unable to determine an accurate phenylalanine 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 has not soaked through to the back of the card

Chapter 5

Blood samples

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.
The sample can be taken from any finger, but is best taken from the middle and fourth fingers. Generally the sides of the fingertip are better to use than the pad.

'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.'

- card has got wet
- foot or finger has been squeezed
- card has been placed on a surface while blood still wet
- card has not been allowed to dry slowly in room air
- card has been placed in a plastic bag or container.

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 to flow.

Involving children in their blood tests

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

How to involve your child:

- let toddlers decide which finger to use for the blood sample
- 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

- 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 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.

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 tests sent?

New Zealand
National Testing Centre
P. O. Box 872
Auckland.

Australia
Newborn screening services in Australia are provided by five centralised screening laboratories. These are Western Australia; South Australia that also covers part of Northern Territory; Victoria that also covers Tasmania; 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 mailed to people with PKU.

Chapter 5

Blood samples

What affects phenylalanine levels?

It is normal for Phe levels to vary a little throughout the day. That is why there is an acceptable range of Phe levels rather than one number. Levels are generally higher in the morning because, after not having eaten or taken supplement overnight, 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 drink of supplement.

A number of factors cause blood Phe to rise or fall to a level outside the acceptable range, including the following:

- ↑ Phe (protein) intake from food is too high – which causes the Phe level in the blood to rise
- ↑ a child's rate of growth has slowed and they may be having more Phe/protein from food than they need
- ↑ not eating enough food or getting enough supplement for growth – in this situation the body breaks down its own tissues (e.g. muscle), releasing Phe into the blood and causing the levels to rise
- ↑ sickness – when it may be difficult to eat or take the supplement
- ↑ weight loss – it is best to lose weight gradually to prevent high blood Phe levels (see Losing Weight On The PKU Diet in chapter 10).
- ↓ not eating enough phenylalanine/protein from food will result in low Phe levels
- ↓ weight gain (adult) or rapid growth (child) also causes levels to fall as building new tissue uses up Phe.

If your levels, or your child's, are too high or low your clinic team will discuss ways to bring them under control.

Chapter 6

Phenylalanine in the PKU diet

- the PKU diet
- how is the Phe intake counted?
- how to read food labels
- how to calculate protein
- confusing nutrition information labels
- artificial sweeteners
- medications

Chapter 6

Phenylalanine in the PKU diet

The PKU diet

People with PKU get some of the protein they need from the formula or supplement. The remaining protein – which includes the essential amount of phenylalanine the body needs for growth and functioning – comes from food. The PKU diet consists of:

- cereal based foods – e.g. breakfast cereals, crackers and biscuits
- fruit and vegetables
- low protein foods – e.g. special flour, bread and pasta
- fat and sugar.

Foods such red meat, chicken, fish, eggs, milk, yoghurt, cheese, nuts and legumes (e.g. lentils, chick peas, kidney beans) are too high in protein to include in a PKU diet, except when the dietary restriction required is minimal. (See the diagram, Five Food Groups: PKU and Non-PKU, in chapter 10. This compares a PKU diet and a non-PKU diet.)

The amount of phenylalanine needed and tolerated by each person with PKU is quite different. The diet is adjusted according to blood Phe levels and is likely to vary from time to time.

How is the phenylalanine (Phe) intake counted?

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

To do this, you will need a PHENYLALANINE OR PROTEIN FOOD EXCHANGE LIST, which your dietitian will provide. This shows how much protein common foods contain, allowing you to work out how much of the daily protein quota you (or your child) will eat at each meal. It also lists 'free foods', i.e. foods that contain little or no protein, which don't need to be counted.

There are a number of systems used to count protein. Some systems measure the protein in 'units', others measure it in 'exchanges' and others use 'grams'.

For example:

- $^1/_4$ cup green peas = 6 units OR 2 grams OR 2 exchanges
- 1 cup hot potato chips = 10 units OR 3 grams OR 3 exchanges.

New Zealand uses an exchange system (1 exchange = 50 mg). Australia has traditionally used units (1 unit = 15 mg phenylalanine), although a number of clinics have introduced a system of counting grams of protein for some people with PKU. In most foods, 1 g protein contains 50 mg of phenylalanine.

Phenylalanine in the PKU diet

All these systems can be effective for controlling blood Phe levels. Your PKU dietitian will explain the method used in your centre.

How to read food labels

Once you know how to read labels, you can make your own choices from foods in the supermarket. The ***nutrition information panel*** on the food label contains all the information you need to calculate the units/grams/exchanges. When you know how to read food labels, working out the units/grams/exchanges is straightforward.

Nutrition information table

One of the things you need to check when choosing food for a PKU diet is the nutrition information table. This tells you the amount of energy (kilojoules/calories), protein, fat, carbohydrate, fibre and other nutrients the food contains.

The table also tells you how much protein the product contains per serve and per 100 g. From this you can calculate units/grams/exchanges. You can also compare different products and choose the one with the least protein.

Some low protein foods may not need to be counted, but this depends on the way you count phenylalanine or protein. Be guided by your dietitian about what you can count as 'free'.

How to calculate protein

Breakfast rice pops

Nutrition information

Servings per package: 20
Serving size: 30 g (3/4 metric cup)

	Quantity per per Serving	*Quantity per 100 g*
Energy	*460 kJ 110 Cal*	*1530 kJ 367 Cal*
Protein	*1.4 g*	*4.6 g*
Fat, total	*< 0.1 g*	*< 0.2 g*
– saturated	*< 0.1 g*	*< 0.1 g*
Carbohydrate	*25 g*	*84 g*
– sugars	*10 g*	*33 g*
Sodium	*150 mg*	*500 mg*

Phenylalanine in the PKU diet

Calculating Phenylalanine exchanges: New Zealand	*Calculating Phenylalanine units: Australia*
1 exchange = 1 g protein = 50 mg phenylalanine *20 g rice pops contains 0.92 g protein* *Count this as 1 exchange*	*1 unit = 15 mg phenylalanine* *To calculate the units, multiply the protein content by 3* *20 g rice pops contains 0.92 g protein* *20 g = 0.92 g x 3 = 2.76 units* *Count this as 3 units*

To calculate the amount of protein in 20 g rice pops:

Use a calculator!

100 g Rice Pops = 4.6 g protein
20 g Rice Pops = $^{20}/_{100}$ x 4.6 = 0.92 g protein

Confusing nutrition information tables

Working out the information you need from nutrition labels is not always straightforward. When in doubt talk to your dietitian. Here are some examples of labels that can cause confusion:

1. Protein content per serve is <1 g (less than 1 g): When foods contain less than 1 g protein per serve it can be difficult to work out the exact amount of protein in the food. Other labels cause confusion when they tell you the ***weight of a serving size***, when what you want to know is the ***value in one biscuit***.

Corn cakes

Nutrition information

Servings per package: 24
Serving size: 8 g

	Quantity per Serving	*Quantity per 100g*
Energy	*111 kJ*	*1385 kJ*
	26 Cal	*330 Cal*
Protein	*LESS THAN 1 g*	*6.5 g*
Fat, total	*LESS THAN 1 g*	*8 g*
- saturated	*LESS THAN 1 g*	*1 g*
Carbohydrate	*6 g*	*75 g*
- sugars	*LESS THAN 1 g*	*4 g*
Sodium	*60 mg*	*70 mg*

Pack size = 192 g
To work out the weight of one cake, use the information on the front of

the packet. The total weight of the packet is 192 g and it contains 24 cakes. Using your calculator again (to divide 192 by 24) you can work out that if 24 cakes weigh 192 g, then one cake weighs 8 g (one serve in the nutrition table).

So now you know that one cake contains <1 g protein, but is it 0.1 g or 0.8 g? To find out the actual amount of protein it contains, work back from the 100 g value.

100 g product contains 6.5 g protein
1 g product contains 100/6.5 g = 0.065 g protein
1 cake (8 g) contains 8 x 0.065 g = 0.52 g protein

Depending on how you count protein or phenylalanine, products with < 1 g protein may not need to be counted. Discuss this with your dietitian.

2. **Products that give values for the product mixed with water**: If you don't read the nutrition information table carefully it's easy to be fooled into thinking the food is much lower in protein than it actually is, as the following example demonstrates.

Baby rice

Nutrition information

Serving size: 45 g (5 g cereal prepared with 40 ml water)

	Quantity per serving prepared cereal	*Quantity per 100 g prepared cereal*
Energy	*80 kJ*	*170 kJ*
	18 Cal	*41 Cal*
Protein	*0.4 g*	*0.9 g*
Fat, total	*0.1 g*	*0.3 g*
- saturated	*0 g*	*0 g*
Carbohydrate	*2.2 g*	*9.2 g*
Sodium	*0.5 mg*	*1 mg*

A 45 g serving of baby rice is actually 5 g of the dry rice cereal and 40 ml water. All the protein, of course, comes from the dry rice cereal, so if you want to calculate the amount of protein in a larger or smaller serving, remember that 5 g dry cereal contains 0.4 g protein.

Phenylalanine in the PKU diet

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 *ingredient list* on the food label.

Additives 950, 951, 961 and 962 (also called Nutrasweet, Equal, Canderal, Neotame or aspartame-acesulphame) contain aspartame, which is a source of Phe. Another way to find out whether a product contains these artificial sweeteners is to check for a warning statement saying that the product contains phenylalanine. These sweeteners are used mainly in diet drinks, sugar free lollies and chewing gum and some medications.

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

Diet drink

Nutrition information

Servings per package: 3
Serving size: 200 mL

	Quantity per Serving	*Quantity per 100 g (or 100 ml)*
Energy	*4 kJ*	*2 kJ*
	1 Cal	*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*

INGREDIENTS

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

This drink contains very little protein but it does contain phenylalanine so should be avoided by people on a PKU diet. You may think that if a drink contains aspartame, the nutrition information table would include it in the protein. But aspartame isn't actually a protein, so a diet drink can have little or no protein, but quite a lot of Phe in the aspartame. So, along with the nutrition label, you need to check for the warning.

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 send a completed filter paper (newborn screening card) to your testing centre and advise your PKU clinic.
When buying over-the-counter medications always find out if they contain aspartame or aspartame-acesulphame.

Chapter 7

Feeding your child

- when to start solid foods
- introducing solids – three steps
- how to develop good eating habits
- feeding your child from one year onwards
- feeding problems
- how to minimise tension at mealtimes

Chapter 7

Feeding your child

When to start solid foods

Babies are ready to start solid foods when they are able to sit, with support, and hold their head upright and steady. At this time, they may also be starting to show signs of being interested in what their parents are eating.

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.

Learning to eat solid foods generally takes babies several months as they get used to new tastes and textures.

Your PKU 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.

Counting protein

As the variety of foods in your baby's diet increases you will learn about counting protein in their food. (See chapter 6). This is not difficult but needs to be done accurately.

Foods are measured and the amount of protein your child eats is added up using the PKU foods list – your dietitian will give you this.

It's a good idea to write down the foods your baby has eaten at first, 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. (see Sam's Phe Planner in Chapter 8.)

The low phenylalanine formula will always be needed. The recipe, amount and type of formula will change as your child gets older.

How to give solid foods to your baby

Choose a quiet time of day when you and your baby are relaxed after a breast or formula feed. In a quiet place, seat your baby in a secure position facing you. Use a small spoon with smooth edges. It is not necessary to add solids to your baby's bottle.

Place a small amount of food on the spoon and hold it to your baby's mouth. Press the spoon gently down on your baby's lips. When their mouth opens, place the tip of the spoon just inside. Babies take the food from the spoon in a sucking motion.

At first, babies 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. Your baby will gradually learn to take the food off the spoon and swallow it. Eating skills improve quickly over a week or two.

All babies have their own ways and preferences where food is concerned. Give your baby solids that suit their feeding skills.

Introducing solids – three steps

Solid food is introduced in three stages:
1. smooth and lump free food
2. lumps and chunks that can be chewed
3. self feeding and finger foods.

Step 1: smooth and lump free food

Fruits and vegetables are mostly low in phenylalanine, and make good first foods for your baby with PKU. Smooth, lump free foods – foods that are sieved or pureed – can be made at home, or bought ready made as jars or cans of commercial baby food. Strained fruits, fruit gels or vegetables are suitable.

Start with a small amount, 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 once or twice a week.

Suggested first foods include:
- strained (pureed) vegetables such as pumpkin, carrot and green beans
- baby fruit gels
- strained (pureed) fruits such as 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.

Step 2: soft lumps and chunks

At around eight to nine months babies start to make chewing movements, whether or not they have teeth. This is the time to introduce thicker and lumpier foods. You can now give food **before** a breast or formula feed. Also at this stage:

- foods can be mashed, grated, diced and pureed more thickly than before
- you can increase the variety and amount of foods your baby eats – try mixtures of vegetables, vegetable soup, fruit and cereal, low protein pasta and vegetables, baby cereals and low protein rice (see PKU recipe books

for ideas)
- offer sips of water from a sipper cup
- keep giving Phe-free formula from a bottle, or try tiny amounts from a sipper cup
- your dietitian will show you how to count the protein in your baby's food.

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

Meal planning

As new foods are added, the task of meal planning begins:
- aim to spread the number of units/grams/exchanges throughout the day
- decide how many units/grams/exchanges you have for that meal or snack
- choose the foods you will offer
- choose one higher Phe food such as cereal, potatoes, peas, or corn
- choose a lower Phe vegetable or fruit such as carrots, green beans, peaches or pears
- measure the amount of food your child may have to add up to the number of units/grams/exchanges 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 units/grams/exchanges
- as your child eats more foods, they will need less of their regular formula
- be sure to let the dietitian know if your child is not eating all of the units/grams/exchanges – the formula may need to be adjusted.

Step 3: self feeding and finger foods

Once babies are managing lumpy foods they start to become interested in feeding themselves. This usually happens at around 10 to 12 months of age, though you can start offering finger foods from seven months onwards.

Babies will be eager to help feed themselves with fingers and a spoon if given the chance.

Learning to eat is a messy business! Food will be dropped and spilt. Easy-to-clean baby equipment makes life easier.

If babies get used to sitting in a highchair or at a low table when eating, it keeps them focussed on eating and also helps to confine the mess. Placing a plastic mat on the floor makes spills easier to clean up.

Ideal finger foods include:
- hard dry toast made from low protein bread or PKU rusks (see Recipes in chapter 10)
- peeled soft fruits
- strips or pieces of cooked vegetables
- low protein pasta spirals
- low protein crackers

When feeding your baby from a bowl use two spoons, one for them to practice with and one for you. Offer soft cooked table foods cut into small pieces at 10 to 12 months.

Try adding herbs and spices to make food tastier. Remember that your baby will be still enjoying the natural tastes of foods and there is no need to add salt.

Begin to offer some formula in a sipper cup and work towards weaning from the bottle by one year of age.

'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.'

How to develop good eating habits

The following suggestions may be helpful:
- establish a meal and snack schedule
- offer food at the table or in a high chair
- discourage eating between meals and snack times
- offer formula at meal times, and water as the other drink
- be aware that changes in appetite and intake are normal
- continue to offer reasonable choices at consistent times, even though your child may sometimes refuse food or become 'picky'
- have your child sit at mealtimes whether or not they are eating
- stay positive about food and formula and avoid power struggles.

Mealtime, family time

Your child will want to have foods they can't eat. You will need to tell your child they are special and show them which foods they can have. The clinic team will provide tips on how to avoid or manage mealtime problems with siblings and extended family.

Include your baby in family mealtimes from an early age. This will encourage good eating habits and help your child to learn that their diet is special.

Feeding your child

Phenylalanine-free formula

As your baby eats more solid food and less breast milk or infant formula, there will be changes with the phenylalanine formula (called the 'supplement' after your child is 12 months of age). The supplement is still essential, but your dietitian will change the recipe, and the type of supplement (read more about phenylalanine-free supplements in chapter 4).

The supplement replaces high protein foods such as meat, fish, chicken, eggs and cheese, and will need to be taken every day as directed by your dietitian.

'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 juice for a treat once or twice a week, and limit it to 100 ml a day. Too much juice dulls a child's appetite for food, may make them overweight and isn't good for their teeth.

You may talk about foods as: 'yes, no, or maybe'. 'Yes' foods are free and very low protein, such as apple, carrots and low protein foods. 'Maybe' foods must be measured, such as crackers, rice, pasta, 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 grains, cereals, bread, pasta and biscuits. Here are some other suggestions:

- offer small pieces of suitable soft-cooked family foods
- give most fluids by cup
- have set meal and snack times
- count the protein carefully
- you may start offering a set number of units/grams/exchanges at each meal and snack
- continue to keep a record of units/grams/exchanges – 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
- if you child is still drinking standard infant formula, cow's milk may be substituted

- the amount of Phe-free formula your child drinks should be about the same every day, as prescribed
- your child will be having their supplement every day
- start introducing more low protein foods.

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.

Feeding problems

Refusing food 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.

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 spoon-fed. Give them their own spoon to try. Offering plenty of finger foods encourages independence.

How to minimise tension at meal times: 7 tips for success

1. Respect that your child may not be hungry.
After the age of 12 months children don't grow as quickly, which means their appetite won't be as large.
2. Watch for 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. It can happen especially when you have made a lot of effort to prepare meals for your child with PKU. Some children refuse to eat, knowing it is an effective way to gain attention.

Remember to stay calm if your child refuses their meal. There may be an opportunity to make up some units/grams/exchanges later or the next day, or your child may just have a low day. The blood test results will tell you if there is a problem.

Chapter 7

Feeding your child

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.

Aim to have three meals with a snack mid morning and mid afternoon. Don't let your child eat constantly throughout the day. Let them know that it will soon be time to eat.

'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 PKU team suggested I give him his food before his supplement when he was really hungry. I let him get down from the high-chair 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 with the spout.'

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. Encourage all the family to eat plenty of fruit and vegetables – as a good example, and for good health.

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 PKU clinic team or other parents about approaches they have found helpful.

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

Try to keep the mealtime atmosphere relaxed so that this is an enjoyable time for the family. Turn off the television and try to avoid having family arguments 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.

Seat your child at the table for meals. Children have short attention spans. Set aside 20–30 minutes for meals, and 10–15 minutes for snacks. Forcing your child to sit for longer may lead to further refusing of food.

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.

7. Keep a positive attitude to your child's diet.
Allow your child to form their own opinions about their diet. Stay calm, or avoid showing your anxiety – which will reduce the tension.

Feeding toddlers

It can be reassuring to remember that:
- most healthy children will not starve themselves
- it is quite normal for children's appetites to vary from one day to the next
- if they are growing well, they are eating enough
- when feeding toddlers, expect waste and mess
- the clinic team is there to help if you are having trouble with feeding.

Chapter 8

Encouraging independence: talking about PKU with your child

- encouraging independence
- what to expect of your child at different stages
- talking about PKU with your child
- daily Phe planner

Chapter 8

Encouraging independence: talking about PKU with your child

Encouraging independence

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

Fostering independence in managing their PKU has a number of advantages. It:

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

By allowing your child to take charge as they mature, some of the more difficult aspects of their treatment, such as taking blood and diet restrictions, become a shared responsibility. It also means they will be less likely to blame you for their treatment.

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.

What to expect of your child at different stages

Toddlers: 2–5 years

- are aware that they have a special diet
- know they need to take their supplement
- know they need to have blood tests
- know to check new foods with their parents
- watch you prepare the supplement
- start to learn yes/no foods
- are aware that the Phe in food is counted.

Early childhood: 5–8 years

- understand that they have a special condition called PKU
- understand that they can't eat high protein foods
- have a basic knowledge of why the supplement is important
- know what is considered a high Phe level

- help you prepare the supplement
- practice using scales to weigh foods and count Phe
- have ideas about meal choices
- learn to select low protein foods in social situations
- can assist in taking their own blood samples.

Late childhood: 8–12 years

- are able to prepare their own supplement
- can weigh foods accurately and count the units/grams/exchanges
- are able to make appropriate meal choices
- are able to take their own blood samples
- have a basic understanding of PKU, Phe levels and low protein diet
- know how frequently they need to take blood samples
- understand that the supplement helps provide nutrition and lowers Phe levels
- can read and calculate the units/grams/exchanges from product labels with assistance.

Early teens: 12–14 years

- prepare their own supplement all the time
- can measure Phe accurately
- are able to make appropriate meal choices
- can prepare some basic meals for themselves
- take their own blood samples
- have an understanding of what causes PKU, phenylalanine, enzymes, amino acids and the effects of illness
- know the healthy range for blood Phe levels
- understand risks and effects of high Phe levels
- make an effort to expand low protein diet choices
- are able to keep a food diary to record protein intake
- read and calculate the units/grams/exchanges from product labels without assistance
- are aware of the pregnancy risks associated with maternal PKU
- attend an adolescent PKU clinic or have a plan in place for transition to an adult metabolic service.

Late teens: 14–18 years

- prepare their own supplement
- can measure Phe accurately
- are able to make appropriate meal choices
- can prepare PKU meals and non-PKU meals
- remember to take their own blood samples
- send blood samples for testing

Chapter 8

Encouraging independence: talking about PKU with your child

- are able to make dietary changes depending on blood Phe levels
- 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
- know how often clinic visits are needed and take responsibility for making their own appointments
- know how to contact their consultant, dietitian or nurse when required
- are aware of the pregnancy risks associated with maternal PKU
- attend an adolescent PKU clinic or have a plan in place for transition to an adult metabolic service.

'Mum always used to say to me if you eat the wrong foods you won't grow. It's up to you, but if you want to be short for the rest of your life . . . That's the only way I could understand it at that age, I didn't understand brain damage, I just used to think I'd better eat this because I want to be tall.'

Talking about PKU with your child

Talking to your child about PKU and its day-to-day management is beneficial even at an early age. The PKU clinic team will support you in this, answering any questions you may have, and helping you increase your child's knowledge and independence.

By the time children start school or have meals at friends' houses they need to have an understanding of their diet and the foods they are allowed to eat. Help your child to make decisions about aspects of their diet – such as taking their supplement to school, or whether to take food to a special party or sleepover, or choose food they are allowed from what is provided.

Having PKU, they will be asked questions about their diet.

Frequently asked questions include:
- Why don't you eat meat?
- Are you vegetarian?
- Are you sick?
- Is it contagious?
- How can you drink your supplement, it smells?

Here are some possible answers:
- I'm on a special diet.
- I don't mind not eating meat – I'm used to not having it.
- No, I'm not sick – I'm healthy and my diet keeps me healthy.
- It's no more contagious than vegetarianism.
- You can't catch PKU, you have to be born with it.
- My diet is like a vegan diet but even stricter.
- I have taken a supplement since I was a baby, so I'm used to it.

Encouraging independence: talking about PKU with your child

- I'm used to having the supplement. It's like medicine – if you need to take it to be healthy, you take it.

Rehearsing answers and scenarios with your child beforehand may help them avoid being tongue-tied or embarrassed when they are confronted with questions.

Games

Regular discussions about PKU with your child will help them feel more comfortable talking about PKU, and give them the confidence to ask you questions and share their concerns. You can help your child to understand PKU in a variety of ways from an early age. Many families use games, such as the following, or invent their own:

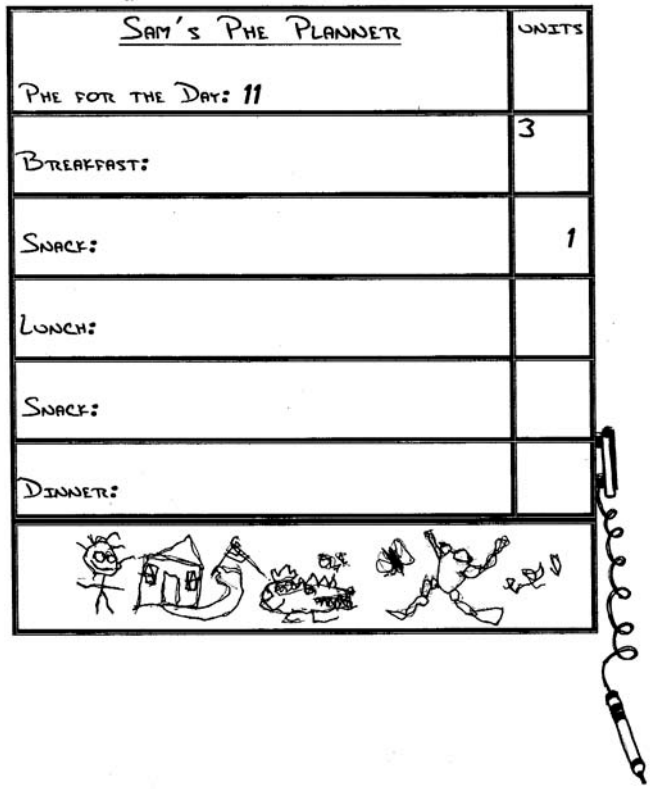
- placing different foods into 'yes' and 'no' categories
- cutting out different foods from magazines and pasting them into a PKU scrapbook
- letting your child rate new foods or meals from 1–5 to encourage them to try new foods
- putting their Phe results on a wall chart so they can monitor their own progress
- cooking PKU recipes to take to school or share with the family
- a star chart/reward system for taking blood tests, drinking their supplement and managing their diet.

The more PKU can become just another fact of life, rather than a burden or an embarrassment, the easier their life and yours will be.

Encouraging independence: talking about PKU with your child

Daily Phe planner

Why not count the daily units/grams/exchanges together? Attach a magnetic board to the wall and write in the amount of the daily allowance you will offer at each meal. For each unit/gram/exchange, place a magnet on the board. As the units/grams/exchanges are eaten during the day, your child removes magnets from the board until they are all gone. Another option is to laminate the chart and stick it to the fridge. Write on it with a whiteboard marker and keep a tally that way.



Chapter 9

Preparing for childcare, preschool and school

- preparing for childcare, preschool and school
- discussing PKU with the school
- food at school
- taking the protein supplement
- lunches, parties and cooking days
- what to discuss with your child

Chapter 9

Preparing for childcare, preschool and school

Pre-planning

This is a time of anticipation and, if it is the first time your child will spend long periods of time out of your care, some concern.

With pre-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, school and the PKU team.

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

Discussing PKU with the school

Who do you need to inform?

It is important that teachers and others at childcare, preschool or school understand why your child needs a special diet, and why it 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 after school care
- the tuckshop or canteen supervisor, or cook.

What to discuss with staff

Firstly, they will need a basic understanding of PKU and the importance of the low protein diet. This may be the most difficult part of the discussion. You are probably familiar with the terminology and concepts of PKU, but explaining it simply to others is sometimes difficult.

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 phenylalanine (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 developing brain
- staying on a protein restricted diet keeps the Phe levels in a safe range and the child will develop normally
- eating the wrong foods will not make them immediately sick, but will have a detrimental effect long-term
- a child with PKU has a very specific diet that is calculated by a specialist

team, with portions measured out daily by the child's family – so it's important that there is a system in place at school to supervise the type and amount of food they eat during the day
- parents must be informed if the child has eaten food that is not allowed, or does not eat foods that are sent from home.

At the end of this handbook, you'll find sample letters which offer a simple explanation of PKU for preschool and school teachers, and also for parents of classmates. You could also photocopy relevant sections of the handbook for them to refer to as needed.

Either you or a member of the PKU clinic team can answer questions and review the main points.

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

Food at school

Each childcare facility 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 at school
- provide the main meals but use the school canteen or preschool meals for snacks, such as fruit, salad plates or ice blocks – you could also give the preschool or school a list of foods that are 'free' (contain minimal or no protein) or allowed in measurable quantities
- use only the school facilities – by either pre-ordering the low protein foods from the preschool menu, or providing low protein bread for the school canteen to make into sandwiches with low protein fillings.

Taking the protein supplement

Your child is likely to be happy to drink the supplement at the usual times at childcare. Label it with their name and store it in the childcare centre refrigerator.

Encourage your child to take the supplement to preschool or school to help spread the intake throughout the day. Carrying it in covered drink bottles or pre-made drink packs (if available) helps avoid questions.

'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.'

'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 I measured him and told him it looked like he wasn't growing. Tears came to his eyes, then he told me himself about the lunch swapping and it stopped.'

Preparing for childcare, preschool and school

Lunches, parties and cooking days

Prepare for these ahead of time by keeping a supply of pre-made low protein cup cakes in the freezer, or low protein biscuits and lollies with your child's teacher.

If the class is learning about food or cooking, your child can share food such as low protein pikelets, so they're not excluded. (See chapter 10 for more ideas about food for special occasions.)

'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.'

School camps or overnight excursions require more planning (see School Camps and Excursions in chapter 10).

What to discuss with your child

Talking to your child is an important part of preparing for 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 appropriately. Things to talk to your child about include:

'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.'

- knowing which foods are OK and which to avoid
- bringing home uneaten food in the lunch box so that you can calculate units/grams/exchanges
- buying only low Phe food from the canteen (and no diet drinks)
- not swapping lunches with friends
- deciding when to drink the supplement
- how to explain the different diet to other kids
- what to do about teasing/embarrassment about the diet.

Children often find it difficult being different from other children, and they feel embarrassed or ashamed about having to eat different food. Offer encouragement to your child by:

- giving positive messages about the yummy food they can have (which may be treat food for non-PKU children)
- role playing responses to comments from other children
- reinforcing to your child that they are special, and that this special way of eating is to keep them healthy and help them to grow
- talking to your child about other people you know who are on a special diet, even if they're an adult, so your child knows they are not the only one on a special diet.

Chapter 10

The PKU diet

- what makes up a PKU meal?
- preparing a PKU meal and family meal together
- PKU meal ideas
- lunch box suggestions
- snacks
- preparing for camps and excursions
- children's birthday parties
- Christmas, Easter and other festivals
- teenage and adult parties
- barbecues
- eating out
- cooking non-PKU meals for family and friends
- fat facts
- controlling your weight
- recipes
- cookbooks
- shopping list

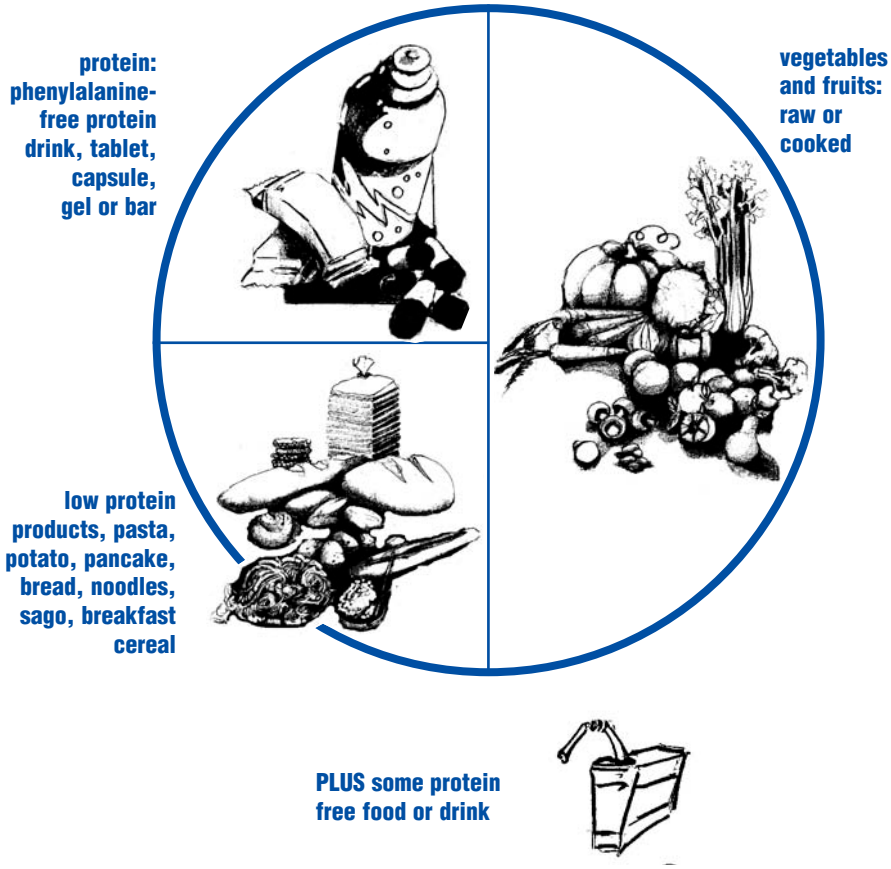
Chapter 10

The PKU diet

Family meal times provide a good opportunity to talk about the day and also for children to learn about healthy food choices. From around a year of age onwards, it's important to start incorporating a child's PKU food options into the family meal setting. This helps them to feel part of normal everyday life.

One way to incorporate the PKU diet into the family meal is to base the low protein PKU meals on the vegetables, cereals or grains that the rest of the family will be eating. The illustration, What Makes Up A PKU Meal, below shows the different parts of a PKU meal. The chart, Family Meal, PKU Meal on the next page has ideas for easy ways to prepare the family meal and the PKU meal together.

What makes up a PKU meal?



Preparing a PKU meal and family meal together

Family food	PKU meal
	Use egg replacer and low protein cheese, and make sure all pasta, rice, flour, sauces and biscuits used in the PKU meal are low protein.
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*
Quiche or pie	*Grated vegetables (zucchini, parsnip, carrot, etc.):* *• baked as a pie with herbs* *• with a low protein pasta sauce stirred through and a low protein bread crumb topping* *• to make vegetable quiche using egg replacer* *• piled into a vol-au-vent shell*
Grilled meat and steamed vegetables	*Fried or baked crumbed zucchini or eggplant slices and steamed vegetables*
Stir fry with rice or noodles (cook the chicken or meat separately under the grill, and use the vegetable stir fry for everyone)	*Stir fry vegetables and low protein rice or pasta with a sauce, e.g. sweet and sour sauce*
Sausages, tomato sauce, mashed potato or chips and salad	*Sandwich mashed herbed vegetables between protein cracker biscuits pre-cut into finger sized pieces, tomato sauce, mashed or chipped parsnip*
Beef patties in hamburger bun or roll	*Vegetable patty in low protein toasted bread*
Tuna mornay or macaroni cheese	*Vegetable macaroni bake: herbs, low protein pasta, low protein cheese and a white sauce using Duocal, rice milk or a stir-through pasta sauce, baked with a low protein bread crumb or sliced tomato topping*

The PKU diet

Family food	PKU meal
Pizza	*Low protein bread (or pizza base made with low protein flour) spread with tomato paste and topped with grilled vegetables and low protein cheese; or low protein bread or cruskits spread thickly with mashed vegetables and herbs, baked till crisp*
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 herbs and oil or a stir-through low protein pasta sauce and salad*

Desserts

Use low protein breads and cereals in the dessert if they were not part of the main PKU 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 with fruit, honey or sugar, lemon juice or cinnamon
- low protein rice or pasta with sugar or honey and margarine, cream, rice milk or Duocal
- low protein rice with fruit and cream
- sago or tapioca with various flavours.

Add the following very low protein foods or free foods to give extra calories and satisfy hunger:

- low protein jelly
- fruit based ices
- vege chips
- low protein custard
- fruit snack packs or fruit gels
- cream.

PKU meal ideas

Low protein foods

As the range of low protein products increases, people with PKU have more options. Snacks and meals that mimic foods from many nationalities are available. With these low Phe foods, Phe allowance can be used more creatively.

'For a quick dessert we purchase meringue nests, fill them with whipped cream from a can and put sprinkles or fruit on top. Yum! We also have Jelly Cups as a snack or for after dinner.'

Mediterranean and Asian

Serve low protein spaghetti or rice with:

- tomato, onion, oregano and garlic stirred through for an Italian flavour
- vegetables, lightly fried and flavoured with soy sauce, for an Asian style meal
- vegetarian sauces.

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.

'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 their noodles and biscuits are low in protein.'

Roasts, burgers, barbecues and more

Try these meal ideas:

- **traditional roast vegetables**, either plain or with flavouring make a good accompaniment to any family meal – serve them in a zucchini 'boat' to add interest
- **quick roast vegetables** – small pieces of onion, capsicum, zucchini, new potatoes, carrot, pumpkin, eggplant, sweet potato tossed in oil and herbs will cook quickly in a hot oven
- **vegetable patties** made with egg replacer and low protein flour mix as binders make an alternative to meat patties – serve with toasted low protein bread and shredded salad to make a burger
- **low protein pizzas** – top with tomato paste, roasted capsicums, mushrooms and onions and bake with a little low protein cheese
- **low protein savoury pancakes** – fill with vegetables and mixed herbs
- **oven-baked jacket potatoes** – halve and scoop out the centre, combine with chopped or mashed cooked vegetables and top with margarine and a sprinkle of paprika
- **potato or sweet potato** – slice, layer and bake with sour cream
- **vegetables** – par-cook, slice and serve cold dipped in oil, salt and garlic or an appropriate dipping sauce, with some low protein bread fingers or crackers thrown in for a great finger meal
- **sweet and sour vegetables** – par-cook and serve hot with a sweet and sour sauce

'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.'

The PKU diet

- **low protein jaffles or toastie pies** – fill with mashed vegetables for a quick and easy meal
- **mildly curried vegetables** – cook with vegetable stock and serve with low protein rice
- **risotto style rice** – use diced vegetables, vegetable stock and flavourings
- **barbecue** – use a combination of par-cooked vegetables and fruits on a skewer and serve with a cold low protein pasta salad.

Lunch box suggestions

low protein sandwich fillings	salads	bread and fruit
■ *avocado*	■ *cherry tomatoes*	■ *fresh fruit (cut up and put into sealed plastic bags)*
■ *grated carrot with sultanas*	■ *cucumber sticks*	■ *individual fruit snack packs, fruit bars or dried fruit*
■ *shredded lettuce*	■ *carrot sticks*	■ *fruit slice*
■ *tomato and cucumber*	■ *stuffed olives*	■ *low protein bread rolls*
■ *beetroot*	■ *sprouts*	■ *low protein crispbreads*
■ *gherkin relish*	■ *celery sticks*	■ *rice cakes*
■ *olive tapenade (paste)*	■ *lightly cooked cauliflower sprigs*	■ *cold pizza slice*
■ *leftover chargrilled vegetables*	■ *low protein pasta or rice salad*	■ *low protein muffins, pikelets, sweet biscuits or cakes*
■ *cold vegeburger*		
■ *honey*		
■ *jam*		

'For me, taking food from home is the easiest way to keep track of my Phe 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.'

Winter favourites

Flick through the soup section of your recipe book for ideas – try potato and leek, mixed vegetable, tomato, carrot or zucchini soups. Serve with low protein toast, cubed and sprinkled on top, or dipped as fingers.

To make a vegetable pie, arrange mixed vegetables in a pie dish, top with mashed potato or low protein pastry, and bake until crisp. Use low protein lasagne sheets to make vegetable lasagne.

Cook and freeze vegetable dishes, soups and sauces into portion sizes ready for a quick meal when you're short of time.

Lunch box suggestions

When you're preparing a lunch box for school or work, remember that variety is the key to enjoyment. Providing your child with an appetising packed lunch is a good way to help prevent the temptation to swap lunch with someone else.

Try these lunch box ideas for a change:

- salads: lightly cooked cold cauliflower sprigs with chopped mint and orange or grapefruit; low protein rice salad with chopped vegetables (raw or lightly cooked) and low protein dressing or mayonnaise; finely shredded cabbage, grated carrot, celery, apple and sultanas with low protein dressing
- different kinds of soup in a thermos
- low protein muffins, fruit snack pack, pasta salad
- low protein cheese sandwiches (using low protein bread) with vegetable sticks in low protein dressing.

Packed meals for people with PKU, ***published by UK support group, The National Society for PKU, has some great ideas for school and work lunches. You can read it on the web at:***

www.nspku.org/Documents/Packed%20Lunches.pdf

'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 units and I'd check it when she got home.'

Snacks

Snacks for after school, between meals, or any time:

- tomato salsa with crackers or toast
- fresh, dried or canned fruit
- frozen fruit pieces e.g. grapes, rockmelon, orange, banana, pawpaw, kiwifruit, peach or apricot
- vegetables with a salsa dip
- popcorn
- vege chips
- fruit ice blocks and icy poles
- Duocal, low protein milk, or rice milk shakes or fruit smoothies
- low protein savoury crackers
- low protein baked goods, such as scones, biscuits, cup cakes or pikelets
- low protein sandwiches
- low protein jelly
- low protein pasta made into cup-a-soup
- low protein snack pot
- potato gems
- low protein breakfast cereal with rice milk.

Camps and excursions

With good planning there's no reason why you or your child can't attend camps and excursions. For a day trip, pack the usual lunch and supplement (and include a favourite treat if appropriate). For more tips from others with PKU on how to prepare for a camping trip, going away with friends or staying the night at a friend's place, see the PKU Quiz.

'I've always had a meeting with the teacher well before the camp. We have had successes and disasters. On the last camp the teacher in charge of her diet wasn't there at meals so she missed out. Next time I will get her to ring me at meal times to be on the safe side.'

Getting organised

How to prepare for an overnight or longer camp:

- discuss your requirements with the camp supervisor, including the need for a protein supplement
- try to obtain copies of the menus so you can help plan your menu for the camp
- pre-weigh and package the protein supplement into separate bags so that it just needs to have water added at the camp (or take your supplement in sachets, capsules or tetrapak, which don't need refrigerating)
- take a supply of low protein bread, pasta, crackers, biscuits and Duobar
- include some free foods for midnight snacks
- give the camp supervisor a clear meal plan.

If you're going camping with friends, plan what you will take in advance so that there will be food you can eat. If someone else is organising the food, tell them what you can and can't eat, if possible. Take plenty of low protein food to help keep your energy levels up. Some ideas are:

- dried fruit, fresh fruit or individual fruit snack packs
- low protein noodles and pasta
- ready made pasta sauces
- low protein cereal – eat it dry
- rice milk
- snack bars
- low protein biscuits and crackers
- vege chips – packed in a plastic container so they don't get crushed
- cherry tomatoes
- rice cakes or corn thins
- lollies (without milk or chocolate).

Children's birthday parties

Before the party

Parents of children with PKU worry that their child will overeat 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 units/grams/exchanges 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 units/grams/exchanges than usual.

Parties at home

Plan the food well in advance, involving your child in choosing the menu. Guests who are toddlers and young children may be quite willing to try special PKU foods. Older children may be more aware of the differences between PKU and regular foods. 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 probably not be appropriate for your child
- include lots of regular foods that are low in phenylalanine units for everyone – include some of your child's favourite PKU foods as well as some regular party favourites for the guests
- 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.

'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.'

Making a PKU birthday cake

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

- use a favourite PKU cake recipe and decorate it with icing and low protein lollies – try cutting the cake to make simple shapes before icing it
- make a low protein ice-cream cake or find a ready made one with low protein content
- 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 or the legs
- use low protein sweet biscuits joined together with whipped cream to make a log, cover with cream and refrigerate overnight – add the decorations at the last minute.

'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.'

'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 jelly beans (which she could eat) and coloured pencils.'

Prizes and lolly bags

Use non-food prizes for games – stickers, pencils, hair accessories, bookmarks, diaries or whatever your child is into at the time – make a good alternative. Low protein lollies can be used as prizes and for the lolly bags, also small packets of chips (including vege chips) or biscuits.

Games and activities

These can help take the focus off food. Here are some ideas:

- arrange for a clown, a special story teller or a face painter to visit
- holding the party in a park with picnic food also takes the emphasis off food and allows for activities such as kite flying
- mini golf, adventure playgrounds, ten pin bowling, roller skating or the beach can work well – as food is less likely to be the most exciting event in the day.

'I have found that people are so willing to accommodate his needs if I let them know what they are.'

Some popular games, such as the chocolate game or feeding one another custard while blindfolded, involve food. Try substituting low protein foods for the regular food in the game, for example Duobar chocolate, PKU custard, and low protein lollies for a treasure hunt.

Parties away from home

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. This gives them plenty of time to accommodate your child's needs.

Letting the host know about the kinds of food your child can eat will be helpful. If they want some specific suggestions, you could photocopy the PKU Party Food Ideas section on page 83, highlighting the foods that are easy to prepare and that other children at the party will also enjoy.

Other ways you can help the host and make sure your child is included are to:

- offer to send a platter of food for the table that your child (and others) can eat, or a special plate of food to be given to your child at the time the food is served – let your child help decide what foods will be included on the special platter (and try to drop the food off before the party so your child doesn't feel self-conscious about eating different food)
- deliver a PKU cup cake to the host before the party so your child can have this when others are having birthday cake – or you could suggest that your child eat some of the lollies on the cake instead of the cake
- tell the host that your child should not have diet soft drinks, diet cordials, milk shakes or flavoured milk – regular cordials and soft drinks are fine.

Ask about the games that are planned, and offer to provide appropriate substitutes for games that involve foods. Tell your child to bring lolly bags home so you can replace any unsuitable lollies with something else.

Fast food parties

If the party is to be held at a fast food chain restaurant let the host know which foods your child can have from the menu (e.g. chips, soft drink). You might like to send along something for dessert.

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 phenylalanine 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.

PKU party food ideas

You may like to give this list to anyone who asks you what party foods they can prepare for your child:

- frozen fruit pieces (strawberries, banana, orange segments, grapes)
- a fresh fruit platter
- strawberries dipped in icing sugar or melted Duobar
- fruit pieces on toothpicks
- fruit mice made from tinned pears with sultanas, cherries and lollies for face and tail
- gelato (check protein content) or Vitari ice-cream (Australia)
- chips
- hot chips
- vege chips
- dips
- plain sweet biscuits with icing made from icing sugar, margarine, water and food colouring
- vegetable sticks with dips
- salad platters
- fruit parfait with low protein jelly – or baby gels from jars
- fruit and cream
- low protein jelly moulds
- Duobar shapes

'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.'

Drinks

- regular cool drinks and cordials
- fruit juice cocktails

The PKU diet

Lollies

Write a list of your favourite low protein lollies here:

'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 units 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 using Loprofin flour and I made custard using custard powder and Coffee Mate.'

Christmas, Easter and other religious festivals

Find special alternatives to the usual foods that are part of your family's or your friends' celebrations. Make a delicious vegetable bake, a fruit and cream dessert (fresh or frozen) or a standout dish that is not part of your everyday fare.

For Christmas, bake some low protein mince pies and low protein Christmas pudding or cake. You can make PKU Easter eggs by filling plastic casings from craft stores, such as Spotlight, with melted Duocal. Try melting Duobar 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.

'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.'

Teenage and adult parties

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

Barbecues

Salads are usually provided at barbecues, but you may need to take some of the following to make sure you will have enough to eat:

- 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
- dips, e.g. guacamole, salsa, eggplant
- low protein crackers.

Eating out

Takeaway

Look for these foods if you're eating takeaway:

- chips*
- apple pie*
- fruit salad
- salad
- soft drink (not diet)
- juices
- mashed potato* and gravy
- hash browns*
- corn
- ice-cream*
- pizza* – ask for tomato and vegetables, no cheese; thin crust is lower in protein; many pizza places offer 'no cheese' pizzas, especially as there are more people with allergies these days.

Many takeaway food outlets label their food with the nutrient content so you can work out the protein content.

Commercial vegetable burgers are **not** low in protein.

*These foods may be too high in protein if you usually have the low protein versions. If need be, rather than starving, have a small serve and be cautious about protein intake over the rest of the day.

Cafes

When you're eating at a café, these foods are suitable:

- salad without red meat or chicken or fish or egg or cheese or tofu or nuts or bread or creamy dressings
- fruit
- fruit salad
- bread/roll – (if you are able to have bread) a sandwich with salad or vegemite or banana or avocado, herb or garlic bread*
- bruschetta*
- chips*
- jacket potato with low protein filling – tomato or guacamole or sour cream and sweet chilli sauce*
- vegetable soup (not based on milk, lentils or other beans, e.g. kidney or white beans or chick peas)
- pasta* with tomato sauce (no cheese – ask for no cheese or cheese separately)
- other pasta sauces that may be OK (check the description on the menu) are: napoletana (tomato with basil), arrabiata (with chilli), primavera (with vegetables)
- vegetable based risotto* – ask for no cheese
- side serve of vegetables.

*These foods may be too high in protein if you usually have the low protein versions. If need be, rather than starving, have a small serve and be cautious about protein intake over the rest of the day.

Restaurants

If you eat regularly at a restaurant or know ahead of time where you are going, you'll often find the restaurant will be happy to cook low protein food. Many Italian restaurants will cook low protein pasta (take your own to the restaurant) and create a low protein sauce to go with it. Here are some ideas about what you can eat in different restaurants:

Australian and New Zealand restaurants

- vegetables or salad based meals, e.g. vegetable stacks, ratatouille
- vegetable based risotto*
- potato as chips, wedges or potato in jacket*
- pasta dishes with sauces based on vegetables, e.g. rocket, garlic, olives, artichokes, sweet potato, pumpkin and semi dried tomatoes – if you take your own low protein pasta the chef may be able to cook that and serve it with one of these sauces*
- dressings or sauces, e.g. gravy or barbecue sauce
- bread/roll/salad wrap*
- fruit, e.g. caramelised figs, compote of fruit (stewed fruit), strawberries, grapes in toffee

Italian restaurants
- pasta with tomato based sauces or sauces listed earlier in this chapter, or sauces with mushrooms and cream – no cheese*
- garlic mushrooms
- vegetarian antipasto – artichokes, olives, tomatoes, eggplant, zucchini and capsicum
- stuffed vegetables* – check there's no meat or cheese
- fruit
- gelato (not dairy-based)

Chinese restaurants
- boiled rice, plain fried rice, noodles*
- vegetable dishes, e.g. stir fried vegetables
- prawn crackers*
- lychees
- fresh fruit

Thai restaurants
- boiled rice, plain fried rice, noodles*
- vegetable dishes, e.g. green or red curry (avoid dishes with nuts, e.g. Panang curry)
- fresh fruit

Indian restaurants
- boiled rice, naan bread, chapatis, papadums, parathas*
- dishes made with sago
- vegetable dishes without legumes (lentils, red or white beans, chick peas)
- mango chutney and pickle, lime pickle

Malaysian restaurants
- boiled rice, plain fried rice*
- vegetable dishes, e.g. stir fried vegetables, avoid vegetable dishes cooked with blachan (fish paste)
- sago based desserts, e.g. gula melaka
- fresh fruit

Greek restaurants
- rice, cracked wheat (bulgur), bread*
- vine leaves*
- dips – eggplant
- olives
- vegetable dishes, e.g. vegetable kebab
- salad

Mexican restaurants

- potato wedges*
- tortilla chips*
- corn chips*, taco shells*
- sour cream, salsa
- guacamole.

*These foods may be too high in protein if you usually have the low protein versions. Have a small serve and be cautious about protein intake for the rest of the day.

Cooking non-PKU meals for family and friends

Adapting PKU meals for a non-PKU diet is simple. You will need to add a source of protein, such as red meat, chicken, fish or cheese. Vegetarian meals are suitable, but will also need to include protein, such as cheese, nuts or legumes (lentils, chick peas, 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.

Food on the run

Eating well on the run is all about getting organised. Having plenty of food supplies in the house makes it easy to throw a few things together in a hurry.
See Lunch Box Suggestions on page 78 for ideas about what to take if you're away from home for the day. If you're buying lunch, choose low protein foods as much as possible. Take plenty of snacks in the car if you're out and about or travelling.

Preparing dishes for the week and freezing them means you can come home to a quick dinner.

Fat facts

Certain fats are good for the body, others should be eaten in moderation. Intake of some of the healthy fats can be low in a PKU diet. Food labels give information about the amount and type of fat the food contains. Try to eat a variety of fats and oils.
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.

Saturated fats tend to raise blood cholesterol levels. In a PKU diet many of the common sources of saturated fats, such as meat and dairy products, are not eaten. However, it is still possible to have a high intake if your diet includes large amounts of the following: hot chips, commercial biscuits,

snack foods, cakes and pastries, cooking margarine, butter or cream.
Monounsaturated fats have a good effect on blood cholesterol levels and are contained in foods such as: olive oil, canola oil, peanut oil, avocados, olives and some margarines.
Omega-6 polyunsaturated fats have a beneficial effect on blood cholesterol levels, and are found in foods such as: oat bran, rice bran, sunflower oil, safflower oil, and polyunsaturated margarine.
Omega-3 polyunsaturated fats are good for your heart and thought to have other health benefits as well. The PKU diet tends to be low in omega-3 fats since fish, seafood, eggs and nuts are usually excluded. Try to include some of the other sources of omega-3 fat in your diet – canola, linseed, mustard seed or walnut oils, and dark green leafy vegetables – or you can take a fish oil supplement. The body needs the right balance of omega-6 and omega-3 fats to function best, so not overdoing the omega-6 sources can also help.

Controlling your weight

Some people on the PKU diet have trouble keeping weight on; others, especially if they don't do much physical activity, have problems keeping their weight in check.

Losing weight on the PKU diet

Tips for weight loss:
1. Avoid crash diets: Weight loss needs to be gradual – no more than half a kilogram a week maximum – or you will start to break down muscle as well as body fat. Since muscle is mainly protein, this will increase your blood Phe level. Gradual changes lead to gradual weight loss, which is more likely to be sustainable than a quick fix approach.
2. Exercise: Find ways to do more exercise as you change your eating habits. Current recommendations are thirty minutes of moderate physical activity, such as brisk walking, swimming, cycling or dancing, on all or most days of the week. Exercise that builds muscle helps to prevent increases in blood Phe levels while you lose weight, and may also increase the amount of phenylalanine you can tolerate in food.
3. Supplement: Take the amount of supplement your dietitian recommended. If you don't take enough you're more likely to break down muscle, which will cause blood Phe levels to rise. Taking more will not cause harm, but since the supplement contains energy (kilojoules), it will make losing weight more difficult. Ask your dietitian's advice on which supplement is best when you're trying to lose weight as some are lower in energy.
4. Have your usual phenylalanine (protein) intake.
5. Eat three meals and three snacks a day so you don't get too hungry. Include vegetables, fruit and carbohydrates, cooked or prepared with no fat or sugar. Choose from the following:
- vegetables – cooked or raw
- potato, sweet potato, corn, peas

- bread – regular or low protein
- breakfast cereal – regular and low protein
- pasta and rice – including low protein varieties
- fruit
- plain biscuits – including low protein biscuits.

6. Limit high fat foods: You need some fat in your diet so do continue to use *a little* oil or margarine. See Fact Facts in this chapter for information on the best types of fat to choose. How to reduce fat:

- spread margarine or butter thinly on bread or biscuits, or go without
- use only a little margarine or oil when cooking – about a teaspoon per serve
- add herbs, spices, lemon juice or vinegar for flavour rather than using fat
- choose low fat snacks such as fruit, vegetables, vegetable soups, sandwiches and salads rather than hot chips
- only eat high fat foods occasionally and in small amounts.

Keep these high fat foods to a minimum

- hot chips
- cream and sour cream
- Duobar
- fried food including fried rice and fried noodles
- mayonnaise and salad dressing
- potato chips and high fat snack foods
- vege chips
- cream filled biscuits
- donuts

7. Decrease sugar in your diet: Artificially sweetened foods can be used as an alternative as long as they don't contain aspartame or aspartame-acesulphame. Suitable sweeteners are sucralose (Splenda), saccharine and acesulphame potassium. (See Artificial Sweeteners in chapter 6.)

Keep these high sugar foods to a minimum

- cakes
- cordial
- soft drinks
- ice-cream and ice blocks
- sugar
- lollies
- jellies
- sweetened desserts
- honey, jam, toppings
- sweet biscuits
- chewing gum

8. Drink water, unsweetened mineral water, soda water, tea or coffee or the OK diet drinks rather than juices, soft drinks and cordial.
9. Avoid fad diets: Fad diets, particularly high protein diets, are unsuitable for someone with PKU. They may:

- be unbalanced nutritionally, especially if they're based on a limited variety of foods
- promise fast weight loss, but the results are generally not sustained, despite quick weight loss initially
- not promote long-term healthy eating habits
- be harmful to your health, e.g. by causing you to lose weight too quickly or raising your blood Phe levels.

10. Beware of weight loss aids or commercial weight loss diets: Commercial weight loss diets are often high in protein. Weight loss aids such as tablets, drinks and patches that promise easy weight loss may be harmful. Generally, they don't work and all you end up losing is your money. Unless they have been prescribed by your doctor, avoid them.

How to gain weight

You need to eat more food, or to add extra fat and sugar to your food, so that you consume more food each day than your body uses.

Tips for weight gain:
1. Supplement: You will need a little more supplement than you need to simply maintain your weight. Take it throughout the day in three or more lots.
2. Protein: Have the amount of protein or phenylalanine recommended by your dietitian.
3. Frequent meals: Eat at least three meals a day and three snacks.
4. Snacks: Fill up on low protein foods such as low protein pasta and rice, low protein bread, low protein biscuits and low protein cereals.
5. High energy food: Add extra sugar and fat to your food, and choose high energy foods – foods that are high in sugar and fat (see Losing Weight on the PKU Diet earlier in this chapter for a list of foods that are high in fat and sugar.)
6. Energy supplements: If you only have a small appetite try adding a fat and carbohydrate supplement such as Duocal (available on prescription) or a carbohydrate supplement such as Polyjoule or Polycose to your protein supplement. Use Duocal on your cereal, as a shake or in low protein custard. Start with 1 level tablespoon a day and build it up gradually to half a cup a day or as recommended by your dietitian. Add Polyjoule or Polycose to cordial or fruit juice.
7. Energy drinks: Instead of drinking water, tea or coffee, have drink soft drinks, sweetened fruit juice or a Duocal shake.

The PKU diet

Recipes

Cooking for someone on a special 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 folder of favourite PKU recipes, and inventing new ones as they gain experience. Be adventurous, experiment, and share your successes with other families.

Here are some basics to get you started cooking the PKU way. The PKU Association of NSW/MDDA/PKU NZ newsletters are a good source of new food ideas and recipes. Ask your dietitian for more tips, recipes and advice about food preparation.

Substitutes for flour, egg and milk

Flour
Most recipes can be made using one of the special low protein flours or cornflour. Use these just as you would plain flour. Add 1 teaspoon baking powder to 125 g low protein flour for cakes. Add $1^1/_2$ teaspoons baking powder to 125 g low protein flour for biscuits and buns.

Egg
Instead of eggs, use an egg replacer such as Loprofin Egg Replacer, Egg Like, or Orgran Egg Replacer.

Milk
Instead of dairy milk, use rice milk, cream (count the protein), margarine mixed with water (100 g margarine and 100 ml water beaten together), coffee whitener or Loprofin milk (count the protein).

Baking low protein products: cooking tips

Cakes
- Sift the flour three times. This incorporates more air, giving a more evenly leavened final product.
- Fold in the flour lightly with a rubber spatula and mix only until it's combined. Beating will result in a tough, rubbery cake.
- Castor sugar works better than ordinary sugar in most cake recipes.
- Cream margarine and sugar together well, until the mixture turns a pale colour and becomes light and fluffy. This incorporates more air.
- Banging spoons or beaters on the sides of the bowl will cause the air you have carefully incorporated to escape.
- Bake the mixture as soon as it is ready.
- Bake on the middle shelf of the oven – unless you have fan bake. Cooking higher or lower in the oven may result in uneven cooking.
- Use cold margarine. If the margarine is too soft, the cake may be too moist.

Bread

- Sift the flour three times. This incorporates more air, giving a more evenly leavened final product with a finer crumb.
- Use castor sugar instead of ordinary sugar.
- When adding flour, mix lightly with a fork to keep the mixture light and airy.
- Avoid banging mixing spoons on the sides of the bowl or the air you have incorporated will escape.
- Bread makers generally produce a successful loaf of bread. If you're having problems, make sure you're measuring the water accurately – you may need to adjust the volume slightly. This can make a big difference.

Pastry

- Chill pastry for approximately 20 minutes before rolling. This helps to keep it together.
- Roll out between two pieces of plastic wrap to stop it splitting, or don't roll it out and simply press the dough evenly into a dish.
- Margarine should be cold and firm for best results; soft margarine makes the pastry harder to work with.

Bread recipes

■ White bread 1

Ingredients
- $3\frac{1}{4}$ cups low protein flour
- 1 tablespoon sugar
- $1\frac{1}{2}$ cups warm water + 3 tablespoons
- 1 tablespoon + 1 teaspoon Metamucil
- $\frac{3}{4}$ teaspoon salt
- $1\frac{3}{4}$ teaspoons yeast (8 g packet)
- 3 tablespoons coffee whitener

Method
1. Combine all ingredients and beat until smooth.
2. Place in a greased tin, smooth top with warm water and leave to rise in a warm place.
3. Bake at 180°C for 30–40 minutes.

Note: If you're using a bread maker, mix the ingredients *before* placing them into the machine.

Try these variations:

Bread rolls

Shape dough into rolls using wet hands. Have water, not flour on the bench or board and rinse hands after shaping each roll. Place each roll into a muffin tin (greased) and cook at 220°C for 15–20 minutes.

Rolls may be glazed with a little oil. Makes approximately 20 rolls.

Bread sticks

Add extra low protein bread mix to make a stiff dough. Roll out with cornflour into long sausages about 1 cm ($^1/_2$ inch) thick and cut into 8 cm (3 inch) lengths. Place on a lightly greased tray and bake at 160°C for 20–30 minutes till crisp throughout. These are great for dipping into soup or for a snack on the run.

Herb bread

Add fried, diced onion, garlic, parsley or other herbs to the dough and mix in well.

White bread 2

Ingredients
- 400 g low protein flour
- $2^1/_2$ tablespoons Metamucil (regular variety)
- $1^1/_2$ teaspoons active dry yeast (8 g)
- 1 tablespoon sugar
- 1 teaspoon salt
- 1 tablespoon vegetable oil
- 380–400 ml warm water

Method
1. Sift flour twice into a bowl.
2. Add Metamucil, sugar, salt and yeast and mix thoroughly.
3. Add dry mixture to bread machine pan.
4. Add the oil and water and start the machine straight away.
5. After 30 seconds or so lift the lid and mix in the flour stuck to the side of the pan. Close the lid. Try to avoid moving or bumping the machine or lifting the lid while the machine is operating.

White bread 3

This recipe is suitable for a smaller bread maker.

Ingredients
- 300 g low protein flour
- 1 teaspoon active dry yeast
- 4 teaspoons Metamucil
- 2 tablespoons coffee whitener
- 1 teaspoon salt
- 280 ml water

Method
1. Sift flour twice into a bowl.
2. Add Metamucil, yeast, coffee whitener and salt and mix thoroughly.
3. Add dry mixture to bread machine pan.
4. Add water and start the machine straight away.
5. After about 30 seconds lift the lid and mix in the flour stuck to the side of the pan. Close the lid. Don't lift the lid while the machine is operating or move or bump the machine.

Flat bread

Ingredients
2 cups (290 g) low protein flour
250 ml water
$^1/_8$–$^1/_4$ teaspoon yellow food colouring
favourite herbs or spices to taste

Method
1. Beat the ingredients together and spread onto well greased greaseproof paper on a greased baking tray.
2. Brush the top lightly with 1 tablespoon oil or water.
3. Bake in a very hot oven for 40 minutes. Remove grease proof paper after cooking.

Rusks

Ingredients
1 cup low protein flour (145 g)
$^1/_3$ cup water

Method
1. Work bread mix and water together to form dough.
2. Roll out with cornflour into a long sausage and cut into 12 lengths.
3. Place onto a lightly greased tray and bake in a moderate oven (180°C) for 20–30 minutes.
4. Cool. If the rusks are still soft, return them to the oven for a few minutes.

Pizza

Base
225 g low protein flour
pinch salt
2 teaspoons baking powder
50 g margarine
150 ml water (approximately)

Topping ingredients:

Basic topping
25 g margarine
1 onion, chopped
pinch mixed herbs
black pepper
OR use commercial tomato based pizza topping or pasta sauce

Mushroom and tomato
50 g mushrooms, sliced
2–3 tomatoes, skinned and sliced

Chargrilled vegies
60 g chargrilled capsicum
60 g chargrilled eggplant
50 g chargrilled zucchini

Potato topping

70 g sliced baked potato
60 g sliced baked sweet potato
60–90 g grated low protein cheese per pizza (optional)

Method

1. Sift together the flour, salt and baking powder.
2. Rub in margarine with fingertips then add enough water to make soft dough.
3. Turn out onto a smooth surface, lightly dusted with flour, and knead until smooth. Roll or press into a 20 cm (8 inch) round and place on a greased baking tray.
4. Spread the basic topping plus one of the other toppings on each pizza. Sprinkle half of the pizzas with cheese and leave the others without cheese. Bake at 180°C for 30–35 minutes until the base is cooked and the topping is browned.
   Serves 4.

Cakes, biscuits and scones

Date scones

Ingredients

$1^2/_3$ cups low protein flour (200 g)
pinch salt
1 teaspoon baking powder
30 g margarine
60 ml cream
20 g castor sugar stirred into 100 ml warm water
30 g chopped dates

Method

1. Preheat oven to 200°C and grease an oven tray.
2. Sift flour, salt and baking powder together into a bowl.
3. Rub margarine in with fingertips.
4. Add cream and water, then chopped dates and mix to a soft dough.
5. Turn onto a floured board and knead lightly.
6. Roll out 1 cm thick and using a small floured cutter (about 4 cm diameter), cut out 26 scones and place them on the tray.
7. Cook for 20 minutes on the middle shelf of the oven.
8. Store in the freezer or in an airtight container in the fridge.

Sponge cake or cup cakes

Ingredients

200 g low protein flour
110 g margarine
110 g castor sugar
2 teaspoons baking powder

2 teaspoons egg replacer
150 ml water
1 teaspoon vanilla essence (or to taste)

Method
1. Preheat oven to 180°C.
2. Place all ingredients in a mixing bowl and beat well for 2 minutes using an electric mixer, processor or wooden spoon, until the mixture is light and fluffy and has a soft, dropping consistency.
3. Place in one 20 cm (8 inch) tin or divide into two greased 18 cm (7 inch) tins.
4. Bake 15 minutes for two sponges or 20–30 minutes for one large cake, or until cake is a light golden colour and springs back when touched.
5. Remove from the tin(s) and cool on a rack.
6. Sandwich together with jam or other filling. Dust with icing sugar. Store in an airtight container.

For cup cakes: place 2 teaspoons of the mixture into each paper patty case and cook for 6 to 8 minutes.

This is a very easy sponge to make. You can add extras (e.g. grated rind and juice $^1/_2$ lemon) for variety. This basic recipe can be used to make special occasion cakes, and baked or steamed sponge puddings.

Sweet biscuits

From Special Recipes ***by Eileen Green, The West Midlands Support Group for Phenylketonuria.***

Ingredients
100 g margarine
100 g castor sugar
1 teaspoon egg replacer
few drops vanilla essence
200 g low protein flour
$1^1/_2$ tablespoons water

Method
1. Preheat oven to 180°C.
2. Cream margarine and sugar until light and fluffy.
3. Mix in vanilla and egg replacer.
4. Gradually add the low protein flour and stir well. Add water and mix to form dough. Use your hands if you find it easier.
5. Lightly dust the bench with low protein flour and knead the dough for a minute or so until it is smooth. Roll out to about 5 mm thick.
6. Cut into shapes using biscuit cutters.

7. Place on a lightly greased oven tray and cook 15–20 minutes or until the biscuits are just turning brown.
8. Cool on a wire rack. These keep well in an airtight container.

Variations:

Lemon biscuits
Replace the vanilla with 2 teaspoons of finely grated lemon rind, and the water with 1 tablespoon of lemon juice. Top with lemon icing.

Sweet flan base
Roll out the dough to fit a flan tin. Prick the dough well and bake in a moderate oven until lightly browned. Fill with raw berries or cooked apple, peach or pear.

Coffee biscuits
Follow the basic recipe for sweet biscuits. Omit the vanilla and dissolve 2 teaspoons of instant coffee into the water before using. The coffee dissolves more easily in warm water, but needs to cool a little before being used in the recipe.

Cinnamon biscuits
Follow the basic recipe for sweet biscuits, adding 1 teaspoon of ground cinnamon to the low protein mix instead of the vanilla essence.

Shrewsbury biscuits
Follow the basic recipe for sweet biscuits, replacing the vanilla essence with 2 teaspoons of finely grated lemon rind and 50 g currants (3 tablespoons).

Jammy dodgers
These are great for children's parties. Follow the basic recipe for sweet biscuits, cut out the dough using a 5 cm (2 inch) round cutter. Use another much smaller cutter to make a hole in the centre of half of them. Bake as usual. When cool, spread the complete rounds with jam, if you are making them for a party, use two different types e.g. apricot and strawberry.
Sieve a little icing sugar onto the ring biscuits, and place them carefully on top of the jam. Press down gently to sandwich them together.

Muffins, pancakes, crumpets and pikelets

Apple muffins

Ingredients
- 1 cup low protein flour
- 3 tablespoons sugar
- 2 teaspoons baking powder
- $^1/_2$ teaspoon baking soda
- $^1/_2$ teaspoon cinnamon
- $^1/_4$ teaspoon salt
- 3 tablespoons vegetable oil
- $^1/_3$ cup water
- $^1/_3$ cup (80 g) apple puree

Method
1. Preheat oven to 200˚C.
2. In a medium mixing bowl, stir together flour, sugar, baking powder, baking soda, cinnamon and salt.
3. Mix together oil, water and apple puree in a measuring cup; add all at once to dry ingredients and mix by hand for 20–30 seconds until smooth (do not use an electric mixer).
4. Spoon batter into six greased 6 cm muffin pans (it tends to stick to cupcake liners). Bake for 15–18 minutes. Makes 6 muffins.

Pancakes

Ingredients
110 g low protein flour
200 ml Duocal
1 teaspoon egg replacer
110 ml water

Method
1. Mix the flour and egg replacer with 3 tablespoons of liquid to form a paste.
2. Gradually work in the remaining liquid to make a smooth batter with pouring consistency.
3. Heat a little oil in a medium-sized frying pan. Pour in a small amount of batter, just enough to cover the base of the pan.
4. Allow to cook for a few minutes and when the underside is done, flip the pancake over to cook the other side.
5. Repeat with the rest of the batter. Makes 4 pancakes. Serving suggestions: Sprinkle with sugar and serve with lemon or orange wedges. Fill with fruit fillings or jam of your choice. Drizzle with golden syrup.

Low protein crumpets

Ingredients
2 cups low protein flour
1 tablespoon baking powder
2 tablespoons custard powder
1 teaspoon sugar
$^1/_2$ teaspoon salt
2 cups warm water

Method
1. Mix the dry ingredients.
2. Add water and mix to a smooth batter (about 2 minutes with electric beaters).
3. Allow to rest for 10–15 minutes.
4. Lightly grease frying pan and heat.
5. Grease egg rings.

6. Half fill rings with batter. When the perforations are showing and the top is dry, turn over to brown the top (this may flatten the perforations).
7. Do not brown them too much to allow for toasting.
8. Leave for 7–10 minutes to cool and dry. Makes about 12.

Quick savoury pikelets

Ingredients
- 1/2 cup low protein flour
- 30 g chopped onions
- 1/4 teaspoon baking soda
- 1 teaspoon chopped parsley
- 100 ml water
- 1/4 teaspoon mixed herbs
- pinch pepper
- oil for frying

Method
1. Sift flour and baking soda into a mixing bowl.
2. Add onion, parsley, mixed herbs and pepper.
3. Gradually add water, beating well to remove lumps.
4. Heat a small amount of oil in a shallow frying pan.
5. Drop mixture by the tablespoon into pan and brown on both sides.
6. Drain on absorbent paper and serve hot or cold. Makes 8.

Custard recipes

Vanilla custard

Ingredients
- 1 1/2 tablespoons sugar (30 g)
- 1 cup water (250 ml)
- 1 tablespoon cornflour (10 g)
- 1 1/2 tablespoons cream (30 ml)
- 2 drops vanilla essence
- 2 drops yellow food colouring

Method
1. Blend cornflour with a small amount of the water to form a paste.
2. Place all remaining ingredients in a saucepan, add corn flour paste and mix well.
3. Cook over a low heat, stirring constantly, until mixture thickens. Serves 2.

Variations:

Blancmange

Use 3/4 cup water only. Follow the same method as for the custard and pour into a wet decorative mould.

Velvet

Substitute sugar with 1 tablespoon of topping or Quik, and leave out the food colouring and vanilla essence. Follow the same method as for custard.

Rich cream and water custard

Ingredients
- $^2/_3$ cup water (165 ml)
- $^1/_3$ cup rich cream (85 ml)
- 1 tablespoon custard powder (10 g)
- 2 teaspoons sugar (10 g)

Method
1. Blend cream, custard powder and sugar together to make a smooth paste.
2. Combine with water and cook over moderate heat, stiring constantly until mixture comes to the boil. Simmer for 1 minute.

Low protein potato balls

Ingredients:
- mashed potato
- cooked low protein rice
- low protein flour
- salt
- pepper
- garlic powder
- parsley
- oil for frying

Method:
1. Combine an equal amount of mashed potato (whipped with water and butter) and low protein rice.
2. Season to taste with salt, pepper, garlic powder and chopped parsley.
3. Bind with low protein flour and shape into balls.
4. Shallow fry in the oil until golden brown.

Cookbooks

Cookbooks catering for people on a PKU diet are few and far between, but many cookbooks include recipes that can be easily adapted for the PKU diet. Most metabolic clinics have an extensive collection of low protein recipes and the web is a good place to browse for inspiration. Recipes from other families – regularly printed in PKU NZ Newsletter, MDDA newsletter and NSW PKU Association Journal – are another great source of ideas.

The PKU diet

Cookbooks with recipes for a low protein or PKU diet

- ***Variety in Life for PKU – a recipe book***, published by SHS International Limited
- ***The Low Protein Collection***, published by SHS Nutrition Services
- SHS recipes on the web: www.shsweb.co.uk/metabolic/pat/ and www.shsweb.co.uk/metabolic/pkubook/
- Other low protein and PKU recipes can be found on the web at: www.cambrookefoods.com/fsRecipes.htm; www.dietspec.com; and www.ener-g.com (not all the low protein products used in these recipes are available in New Zealand and Australia but you can adapt the ideas)
- *Special Recipes*, by Eileen Green, The West Midlands Support Group for Phenylketonuria.

Cookbooks with easily adapted recipes

Many cookbooks include low protein recipes or recipes that can be adapted to make them lower in protein. Cookbooks featuring vegetables, salads, vegetarian, vegan, fruit, potato, rice, pasta and noodles are the most likely to contain suitable recipes.

If weight loss is a priority, low fat cookbooks and those endorsed by Diabetes Australia or New Zealand or the Heart Foundation will contain suitable recipes.

Local Council/Municipal libraries generally hold an extensive collection of cookbooks on their shelves. Children's party cookbooks will give you ideas for decorating your child's birthday cake and presenting other foods.

Recipes and protein content

Recipe books generally don't list the protein content of their recipes, so you'll 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 PKU diet:

- leave out the meat in meat and vegetable dishes
- use a low protein milk substitute for milk, e.g. rice milk, Ducoal, coffee whitener and Loprofin Drink Mix
- where a recipe includes a white sauce, use you own favourite low protein sauce recipe instead
- use egg replacer instead of egg
- instead of cheese in risotto use a small amount of low protein cheese or

stir in a spoonful of coffee whitener or Loprofin Drink Mix powder
- use milk-free margarine or oil instead of butter or margarine containing milk if you usually do so
- use cornflour or low protein flour instead of plain flour
- use low protein pasta or low protein rice instead of regular pasta and rice if you usually do so.

General cookbooks

- *Vegetables*, Liley L, Lownsdowne Publishing Pty Ltd, Sydney 2000
- *Not-so-humble vegetables*, in the Australian Women's Weekly Cookbooks series, Ed Clark P, ACP Publishing Pty Limited 1997
- *Salads simple fast and fresh*, The Australian Women's Weekly, Ed Hammal K, ACP Publishing Pty Limited, Sydney 1999
- *Fresh Vegetables*: A New Approach, by The Sydney Market Authority, Ed Gore M, R A Ramsay and Custom Book Company
- *The essential vegetarian cookbook*, Ed Stephen W, Blayney D, Price J, Murdoch Books, Australia 2004
- *Vegan Cooking Recipes for Beginners*, Batt E, Thorsons, an imprint of Harper Collins Publishers, 2002
- *Verdure! Vegetables the Italian Way*, Lootti E, McRae Books 1997, published in Australia by Simon & Schuster
- *The Great Potato Book*, Fabricant F, Ten Speed Press 2001
- *Risotto: over 80 recipes for a classic dish*, Ingram C, Published by Southwater, an imprint of Anness Publishing Limited 2000
- *Trattoria PASTA*, Werle L, Hodder Headline Australia Pty Limited, Rydalmere 1994
- *Pasta Fresca*, La Place V and Kleiman E, William Morrow, an imprint of Harper Collins Publishers Inc, New York 2001

Shopping List

The PKU shopping list includes regular supermarket items and special low protein products. In Australia these can be ordered directly from the supply company (order forms are available from your dietitian). In NZ, many are available on prescription (which your doctor will provide), others can be ordered directly from the company.

The special low protein products are identified with an asterisk (*). Check the nutrition labels on the packets to find out the protein content of each serving. How much you can eat of these foods will depend on your protein allowance for the day.

Some of the foods listed are available only in New Zealand or only in Australia. Talk with your dietitian about suitable local options.

The PKU diet

Breakfast cereals

- Loprofin Breakfast Cereal Loops*
- Kelloggs Coco Pops
- Kelloggs Fruit Loops
- Kelloggs Honey Crispix
- Kelloggs Rice Bubbles
- Kelloggs Corn Flakes
- Sanitarium Ricies
- Sanitarium Weetbix

Bread

- Loprofin Sliced Loaf*or Loprofin Part-Baked Bread Rolls*
- sliced bread
- bread rolls
- English or fruit muffins

Bread mix and flour

- Loprofin Mix (all purpose baking mix)*
- Pinnacle low phenylalanine bread mix*
- Orgran gluten free flour
- cornflour
- tapioca flour
- plain flour (white or wholemeal)
- arrowroot flour

Pasta, rice and grains

- Aproten pasta – annelini, rigatini, spaghetti, tagliatelle and lasagne* (available from Sharpe Laboratories)
- Loprofin pasta – macaroni*, pasta spirals*, spaghetti*, lasagne
- Orgran Low Protein Rigati*, Orgran corn pasta, Orgran rice pasta
- Loprofin Pot Snack*
- pasta – a variety of shapes and sizes
- instant noodles - wheat and rice
- noodles – rice, wheat, tapioca and bean flour
- cellophane noodles
- fresh rice noodles
- Agluten riso * (Sharpe Laboratories)
- rice – any variety
- couscous, polenta, semolina
- sago (seed tapioca), pearl tapioca

Biscuits and crackers

- Aproten low protein cracker toast*, Aproten crispbreads* (Sharpe Laboratories)
- Loprofin Herb Crackers*, Loprofin Savoury Crakers*
- Loprofin sweet biscuits

- Orgran Rice crispbreads, Orgran corn crispbreads
- Cruskits, Rice cruskits, Cracker bread
- rice cakes
- rice crackers
- Corn Cakes, Corn Thins
- Saos, Paradise Lites
- Jatz, Rice Jatz
- Saladas, Premium

Snack foods

Many packaged snack foods are suitable for a PKU diet. Reading the labels is your best guide. Other PKU support group members and your dietitian will also have suggestions.

Fruit and vegetables

- fresh fruit
- canned fruit – in syrup or natural juice (or snack packs)
- fruit juice – in cans, bottles or packets
- dried fruit – e.g. sultanas, dried apricots, prunes
- fresh potato, sweet potato, canned potatoes
- fresh and frozen corn or peas
- fresh vegetables – e.g. carrots, mushrooms, tomatoes, celery, lettuce, onions
- fresh or frozen cassava
- frozen chips, potato gems and hash browns
- frozen vegetables – e.g. frozen stir-fry vegetables, frozen mixed vegetables
- frozen vegetable fingers, bubble and squeak
- canned vegetables – e.g. asparagus, beans, carrots, tomatoes
- fresh herbs and spices – e.g. garlic, ginger, chilli, basil, parsley, coriander

Ready prepared sauces

- tomato paste, tomato puree, tomato-based pasta sauces
- ready made sauces including pasta sauces and casserole bases – check the nutrition labels on the many varieties available

These sauces contain a maximum of 1.5 g protein per 100 ml:

- Dolmio Traditional Bolognese Sauces, e.g. capsicum and garlic; mushroom; tomato and basil; red wine and Italian herbs
- Dolmio Chunky Bolognese Sauces, e.g. tomato, onion and basil; garden vegetable; mushroom and red wine
- Dolmio Cooking Sauce – Vegetable Ragout and Creamy Milano Chicken
- Kan Tong Stir Fry Sauces – all except for the satay sauces
- Masterfood Recipe Bases – suitable flavours include Creamy Thai Chicken and Maggi Tasty Cheese Sauce Mix.

The PKU diet

Soups

Choose low protein soups (check the nutrition label). Ready prepared soups come in cans, bottles and cartons. Suitable varieties include pumpkin, celery, asparagus, mushroom and vegetable.
For dried packet soups – choose vegetable based soups, e.g. tomato.

Alternatives to dairy products

- coffee whitener
- rice milk
- Loprofin Low Protein Drink Mix*
- Loprofin PKU Milk Tetra Pack*
- Low Protein Cheddar Cheese*
- Low Protein Mozarella Cheese*

Miscellaneous

- egg replacer, e.g. Loprofin Egg White Replacer*, Country Harvest Egg-like egg replacer*
- dried herbs and spices
- stock cubes, e.g. Massel Ultracube Choice Vegetable Stock
- salad dressings
- lemon juice, vinegar
- oil
- milk-free margarine

Chapter 11

PKU: teenagers and adults

- staying on diet
- the PKU diet for teenagers and adults
- exercise and sport
- body image
- alcohol
- safe off diet

Chapter 11

PKU: teenagers and adults

Staying on diet

In the past, the PKU diet was stopped at the end of childhood as it was thought that raised phenylalanine (Phe) levels could not cause further damage to the brain and nervous system. Not much was known about the benefits of continuing the treatment into adulthood.

Recent studies show that staying on diet into adult life is beneficial. Some people with PKU who have gone off their diet and then start it again say that when they are *back on diet* they:

- feel better
- look better
- can think more clearly
- are less moody
- 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 complete assignments more easily
- think it is a must for doing final year school exams
- can work things out better, e.g. when trying to think strategically such as in team sports or playing games or at work.

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

High Phe levels may mean, among other things, that you are not able to make judgments 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 *stop taking the supplement* develop problems such as:

- tremors (the shakes)
- nervous system problems such as behaving inappropriately or having mood swings or being confused about reality – not seeing things as they really are
- stiff or weak legs
- headaches
- nutritional deficiencies which can cause ***severe*** problems, e.g. lack of vitamin B12, iron and calcium
- eczema.

If you stop your diet or supplement then it is essential – an absolute must – to talk to your doctor and dietitian so that you do it in the best possible way with the least amount of side effects and know what you are getting yourself into.

PKU: teenagers and adults

Eating a normal diet is very different from being on the PKU diet. It is unsafe just to avoid high protein foods. See the section on Safe Off Diet at the end of this chapter.

Your PKU diet

The diet for teenagers and adults is generally more flexible. (For more information about all aspects of the PKU diet, see chapter 10.)

The acceptable blood phenylalanine (Phe) level may be higher, except for pregnant women (see PKU and Pregnancy, chapter 12) – discuss this with your team. Teenagers and adults may be able to tolerate more Phe in their diet, and to eat more normal (i.e. not low protein) foods – such as bread, breakfast cereals, rice, corn and potatoes.

Using lower volume supplements, tablets or bars may be more convenient when you have a mobile lifestyle.

Many resources are available to help make staying on diet easier. These include recipe books aimed at adult palates, and new low protein products that taste good and are convenient. For more information about reading food labels, see chapter 6. For a list of recipe books and information about low protein products, see chapter 10.

The PKU diet for teenagers and adults

Guidelines:

- the most important thing is to **take the supplement** – have it two to three times a day with meals so it's spread throughout the day (ideally over 12 hours)
- **count protein** – do this as directed by your PKU team, and eat protein three times a day at meals
- **eat plenty of fruit and vegetables every day**, as well as rice, pasta, bread and crackers – low protein or not, depending on your tolerance to Phe
- make sure you're getting **sufficient vitamins and minerals** – take vitamin, mineral and tyrosine supplements if recommended
- **drink plenty of water**
- eat the **right sorts of fats** and limit your overall fat intake (see Fat Facts, chapter 10)
- choose foods **low in salt**
- consume only **moderate amounts of sugar** and of foods containing added sugar
- prevent excessive weight gain by **being active and eating appropriately**
- if you choose to drink alcohol, **drink it in moderation**
- take care to store and cook food safely.

Chapter 11

PKU: teenagers and adults

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

Exercise and sport

Having PKU does not limit your ability to participate in exercise or sport. Regular physical activity is an important part of a healthy lifestyle.

After exercise your body needs fluid, carbohydrate and protein to recover. Drink plenty of fluid, especially water and eat some carbohydrate foods like bread or pasta (regular or low protein depending on your diet).

The type, intensity and length of time exercising will determine how much protein you require. Taking your PKU supplement as prescribed should provide your body with enough protein. Discuss specific quantities with your dietitian if you're concerned.

'Protein powders' that promise to increase muscle bulk are **not** appropriate for a person with PKU as they will increase your blood Phe levels. Your supplement provides the protein your body needs and helps keep blood Phe in good control. To increase your muscle bulk, ask a qualified trainer to design a resistance training program for you.

Body image

How we see ourselves is called our 'body image'. Having a positive body image does not necessarily mean having a 'good body'. It 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 or be 'cool'.

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 comes from many sources including friends, family, the media, and ourselves. Dieting to lose weight is common, but not everyone sees their body correctly when comparing themselves to friends or to images and models in the media.

PKU: teenagers and adults

We have a choice about how we respond to pressures to have the perfect body:

- we can try to fit the ideal 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, and realise that bodies come in all shapes and sizes, and that as long as we look after ours – then that is the right shape and size for us.

How to create a positive body image

- **Be active**: Regular exercise puts you in a better mood and reduces anxiety. This gives you more confidence in yourself and the way you feel about your body.
- **Focus on what you do for your body each day, not on how others respond to it**: You can't control others' responses to you, but you *can* make healthy choices each day that will leave you feeling good about yourself.
- **Engage in positive activities**: Find time every day for a rewarding activity – do some exercise, go for a swim, play sport, have a walk with a friend, have a long bath . . .
- **Develop a realistic ideal for yourself**: Genetics tells us that only five per cent of the population are genetically programmed to look like today's female models – that leaves 95 per cent of us, so we're in good company!
- **Avoid people who give body shape or weight too much importance.**
- **Examine your own self talk and challenge distorted thinking about your body**: Do you have to be a perfect 10 to have friends? Consider what made you choose your friends and what you like about them. It is unlikely to be their weight or flawless complexion.
- **Develop many sources of self-esteem**: So much makes you unique and special beyond your appearance. Develop listening skills to be a good friend, practice a skill in sport or the arts or enjoy a good book...
- **Develop perspective**: The older you get the less importance people place on judging people by their appearance. It gets better!
- **Judge yourself as a whole person, not just a body**: Create a list of people you admire who have contributed to your life, school, community, and the world. Was their appearance important to their success and accomplishments? If their appearance was not important what was?

This information came from: Kearney-Cooke, A. (2003). *'Helping Adolescents Become Strong Adults'.* A Project of the Partnership for Gender-Specific Medicine at Columbia University, U.S.A.

Chapter 11

PKU: teenagers and adults

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 phenylalanine
- other pre-mixed drinks may contain artificial sweeteners and are required by law to be labelled if they contain phenylalanine
- 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.

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

Just about everybody knows that the legal drinking age in Australia and New Zealand is 18. Sensible drinking guidelines include the following:

- men – no more than 4 standard drinks per day
- women – no more than 2 standard drinks per day
- have two alcohol-free days a week
- excess alcohol can contribute to overweight or obesity – for weight watchers, the recommended maximum is 2 standard drinks a day for men, and one for women
- alcohol slows your response time and this will affect your ability to competently operate machinery and drive a car
- consuming alcohol is not recommended during pregnancy.

How much is 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*	*0.8 g*
3/4 of a 330 ml bottle of alcoholic soda		*depends on ingredients*

Safe off diet

Research shows that controlling brain phenylalanine 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.

PKU: teenagers and adults

If you decide to go off diet, whether it's for a short period, or for the long-term, make sure you do it with the help of your PKU team so you can do it in the best possible way.

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

People with PKU who are off diet are at risk of vitamin B12, iron and calcium deficiencies if they're not *eating normally or taking an appropriate vitamin and mineral supplement*. 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 PKU 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 sometime in the future, though some people with PKU who have gone off diet say they have found going back on diet very difficult.

Remember!

- **don't go off diet without talking to your team**
- attend clinic once a year
- eat normally and check with the dietitian that you are doing so

OR

- take supplement or vitamins and minerals

Starting the diet in adulthood

Some adults who have never been treated for their PKU may also benefit from a PKU diet. Even though many of these adults have significant loss of mental ability, there may be improvements in behaviour and mental processing, and other problems such as depression and anxiety may be reduced.

Returning to diet or commencing diet in adulthood is a significant challenge for many people with PKU. Attending clinic regularly, learning about the diet and monitoring your Phe levels takes commitment. It is important to have support.

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. In chapter 17 you'll find contact details for local support groups, and a number of internet sites that may be helpful.

PKU: teenagers and adults

Pregnancy

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 PKU and Pregnancy, chapter 12.

Chapter 12

PKU and pregnancy

- why planning your pregnancy is essential
- who can I talk to about healthy pregnancy?
- what to expect during your pregnancy
- exercise
- obstetric care
- after birth

Chapter 12

PKU and pregnancy

Why planning your pregnancy is essential

Women with PKU whose blood levels are kept low **before** they become pregnant and **right through the pregnancy** can have normal, healthy babies. Safe phenylalanine levels for women planning pregnancy are **lower than usual acceptable levels**.

Many women do not know they are pregnant until several weeks after they have conceived. High phenylalanine levels during these early weeks could cause heart abnormalities and other serious problems for babies of women with PKU.

Planning your pregnancy, and following a strict diet so that your blood levels are not high when you become pregnant, is the only sure way to prevent PKU related problems during 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 diet and take the supplement as prescribed. Don't delay!

Before you become pregnant, you will need to follow a strict diet and take your supplement *exactly* as prescribed to keep your phenylalanine levels in the safe range. Your PKU team will provide advice about keeping your blood levels safe, and tell you how often you will need blood tests.

Both phenylalanine and tyrosine blood levels are monitored closely throughout pregnancy. Tyrosine supplements may be prescribed if your level is low.

In Australia and New Zealand all pregnant women and women planning pregnancy are advised to take folic acid, one of the B vitamins. The supplement contains enough folic acid for your needs so you won't need to take extra.

At all times in the pregnancy high levels of phenylalanine in your blood may damage your baby.

Diet before or during pregnancy will not alter whether or not your baby will have PKU. That will be coded in the genes and can't be changed.

PKU and pregnancy

Who can I talk to about healthy pregnancy?

Your PKU team will help you plan your pregnancy and offer ongoing support throughout.

What to expect during pregnancy

The first 13 weeks

The amount of phenylalanine 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. The following may help if you're feeling nauseous with morning sickness:

- small amounts of food very often – eating every 15–30 minutes 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
- biscuits, bread, fruit, pasta and cereals help settle your stomach
- drink cordial, sports drinks, ginger ale or cola (not diet) or eat ice and icy poles – if you become dehydrated you will feel worse
- if you are getting up during the night, have something to drink and eat to stop you feeling sick when you wake up
- having your supplement is important, try eating something to settle your stomach 15 minutes before you start your supplement – different flavours and smaller amounts may help.

Also during this time:

- start weekly or twice weekly blood tests for phenylalanine – your clinic will advise how frequently
- weight gain is supposed to be slow, but weight loss is not good for your phenylalanine levels or your baby's growth
- you may require a supplement of DHA (docosahexanoic acid), an omega-3 fat.

Aim to keep your weight steady, gaining just two or three kilograms in total over the first 13 weeks. Try to eat regularly to prevent weight loss, as losing even a little weight at this time can make your blood phenylalanine level higher.

From 14 to 26 weeks

During this time:

- your phenylalanine levels may start to drop as the baby starts to grow much larger – you may be able to start eating more phenylalanine in foods
- expect weight gain of around half a kilogram each week
- you may need to take tyrosine as your baby uses it up for growth.

PKU and pregnancy

From 27 to 40 weeks

During this time:

- you will still be able to increase the amount of phenylalanine in your food – some, but not all, women with PKU will eat three times more phenylalanine in food than they did before the pregnancy, and still keep good blood levels
- keep fit with regular exercise
- weight gain should continue as your pregnancy progresses.

Weight changes during pregnancy

For most women, 10–13 kilograms is a healthy weight gain during pregnancy. From the time the baby starts to grow more rapidly, weight gain of up to half a kilogram each week is common.

After birth, breastfeeding your baby helps your weight (and shape) return to normal. Women who breastfeed for at least six months usually lose more weight.

Exercise during pregnancy

Keeping active during pregnancy helps you to feel healthy. It is common to feel tired at certain stages and a walk or swim 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 or obstetrician
- you may need to include extra snacks to make sure you are having enough calories
- if exercise is causing weight loss, you may need to reduce the amount or intensity for a while.

Obstetric care

Your obstetric care before and during the birth will be the same as for a non-PKU pregnancy and birth. The only difference will be that you are on a PKU diet. Your PKU team will advise your obstetrician, local doctor or midwife about your treatment for PKU during pregnancy.

After birth

Like all babies born in Australia and New Zealand, your baby will be tested for PKU and other problems soon after birth. Your PKU team will advise you and the hospital when the test should be done.

Breastfeeding is ideal for babies. Mothers with PKU *can* breastfeed and it may keep your phenylalanine level a little lower too.

Staying on diet is your choice, but diet for life is recommended. If your baby has PKU, both you and your baby will need a special diet.

Chapter 13

Dental care and PKU

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

Chapter 13

Dental care and PKU

Dental care and PKU

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 inside of the teeth to form holes. **Dental erosion** occurs when strong acids in foods, drinks or supplements dissolve away the outside 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. Because of this they may have sugary foods and acidic drinks more often. 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 acid 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're usually given as a drink, gel or paste several times a day. These artificial combinations of amino acids are highly acidic and can dissolve the surface of the teeth and are also sweetened to improve taste and can cause tooth decay.

How can dental disease be prevented?

1. By toughening the teeth with fluoride:

- fluoride makes teeth tougher and more resistant to decay and using toothpaste is the easiest way to get fluoride onto the teeth – start using a fluoride toothpaste as soon as the teeth appear
- if you live in an area that has a fluoridated water supply (such as a major city) use a low fluoride junior toothpaste – some examples are My First Toothpaste (Colgate), Stages (Oral B) or Milk Teeth (Macleans)
- if you live in an area without fluoridated water, full-strength fluoride toothpaste may be more appropriate – ask a local dental practitioner for advice
- wipe just a smear of toothpaste onto your child's teeth using a face flannel or gauze – it is important that an adult takes responsibility for applying the very small amount of paste to the teeth (for young children who don't have PKU, the fluoride toothpaste is not necessary)

- from one year onwards put a smear of toothpaste on the brush
- from the start of school onwards, a child needs a pea-sized amount of full-strength fluoride toothpaste
- as a child gets older they should brush twice daily – especially last thing at night (avoid night-time sweet drinks or snacks afterwards) and after breakfast
- adults should use a full-strength fluoride toothpaste twice a day – before bed and after breakfast.

2. 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 a feeder cup by the age of 12 months
- encourage your child to 'drink up' rather than sip a drink slowly
- frequent exposure to foods or drinks that contain carbohydrates and sugars increases the risk of decay
- give teeth a rest – aim for three meals a day and only two 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 fizzy drinks, cordials and juice for mealtimes or special occasions
- after meals and snacks try sugar-free chewing gum when your child is old enough (check that it doesn't contain phenylalanine – Wrigley's Children's Gum with Xylitol is a safe choice).

3. Have regular check-ups with a child-friendly dental practitioner:

- early visits help your child get used to seeing a dental practitioner – this can make it a lot easier as they get older
- the dental practitioner needs to know that your child (or you) have PKU and to understand what it is
- give your dental practitioner your dietitian's name so they can discuss your child's (or your) dental health
- ask your dental practitioner for preventive advice and treatment
- your dental practitioner will be able to pick up signs of decay and erosion much earlier than you or your consultant 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 phenylalanine (see Artificial Sweeteners chapter 6). The important thing when eating sugary food is to avoid constant snacking or sipping, give teeth a rest, and to make sure good dental care is maintained.

Chapter 14

Sick days

- what happens to Phe levels during sickness?
- what to do if your baby is sick
- what to do if your young child is sick
- gastroenteritis
- coping with sick days – older children and adults
- medications containing phenylalanine
- oral rehydration fluids

Chapter 14

Sick days

What happens to Phe levels during sickness?

Illness may cause a temporary rise in your phenylalanine levels, but no long-term harm. When you are unwell the body starts to break down its own tissues, releasing Phe into the bloodstream. To prevent this, a high energy, low Phe diet is generally recommended during sickness.

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.

Contact your local doctor if your child is unwell just as you would if they didn't have PKU.

Feeding your sick baby

Maintaining your baby's fluid intake is important. Here are some practical guidelines (not relevant if your baby has gastroenteritis):

- offer feeds more frequently than usual – you may find your baby tolerates half strength formula better than full strength feeds
- 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 child'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 your young child is sick

If your young child is unwell:

- encourage adequate fluid intake by offering drinks every hour or two while your child is awake
- dilute fluids and supplement – this may make it easier for your child to drink them
- don't force feed your child – they may reject the supplement later
- offer foods frequently without forcing – and return to the usual Phe intake as soon as your child's appetite returns
- your dietitian may recommend you increase your child's energy intake and will explain how to do this.

Gastroenteritis

General guidelines for children and adults (not appropriate for babies):

- for the first 12–24 hours offer only oral rehydration solution (ORS) or appropriate clear fluids – offer 100–200 ml every hour or two when your or your child are 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 force feed the supplement or to have the full amount of Phe intake until the appetite returns.

Coping with sick days – older children and adults

For older children and adults the general guidelines for sick days (not gastroenteritis) are the following:

- try to keep up an adequate fluid intake by taking a drink of cordial or juice every hour or two the child/adult is awake
- drinks should include supplement, but in smaller, more frequent amounts – diluting more than usual may make it easier to drink
- offer foods according to the child/adult's appetite without forcing and without aiming to include all the Phe allowance – return to usual Phe intake as appetite returns or as determined by blood results.

Medications

All medications prescribed by your doctor are suitable, but if possible avoid those with the artificial sweeteners aspartame, nutraweet, neotame or additives 950, 951, 961 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. Over-the-counter medicines should also be checked for artificial sweeteners.

Oral rehydration fluids

A number of oral rehydration fluids are available from your pharmacy. Some are sweetened with aspartame, nutrasweet, neotame, or additives numbers 950, 951, 961 and 962 and are not ideal 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 for the PKU warning.

Which oral rehydration fluids are suitable for children with PKU?

The following commercial products need to be diluted according to the instructions on the packet:

- Gastrolyte effervescent blackcurrent tablets (not Gastrolyte sachets or Gastrolyte R) (Aventis Pharm Pty Ltd)
- Pedialyte raspberry sachets (Abbott)
- Hydralyte iceblock and ready made liquid available (Splenda)

Sick days

If commercial oral rehydration fluids are not available, or your child refuses to drink them, make up one of the following:

- Dilute cordial: use 10 ml + 150 ml water
- Ribena: use 10 ml + 190 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 sugar free or low joule or 'diet' cordials or soft drinks.

Chapter 15

Overseas travel

- covering letter
- organising supplies for your trip
- lost luggage
- food for the flight
- travellers' tips

Chapter 15

Overseas travel

For people with PKU, 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

If you're travelling overseas always take a customs declaration letter from your doctor, (see the sample declaration form in chapter 18 of this handbook).

The covering letter should list the name and quantity of all the PKU products you will be carrying in your luggage, say 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.

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

Sealed containers

Keep the formula or supplement in its ***original sealed containers*** (packages, cans or sachets) and take a copy of the prescription with you when you're travelling overseas.

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, pharmacists, Medicare offices or online at www.hic.gov.au

Organising supplies for your trip

For longer periods of travel, you will need an additional script from your doctor so you can obtain extra supplies of the 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 all 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 main luggage.

Food for the flight

Contact the airlines beforehand to organise food 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 for the flight and to cover the possibility of travel delays.

Overseas travel

Bottled water

Use uncarbonated bottled water to make up the formula or supplement when you're travelling overseas. So 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.

Travellers' tips

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

- you may be able to freight the supplement to your destination beforehand to reduce your luggage
- some people with PKU have posted the supplement to themselves at Poste Restante (to the local post office in a particular city) and collected it when they arrived – if you're posting supplement (or formula) overseas include a copy of the doctor's letter and local contact numbers, if known, in the package
- ask your clinic team to locate a PKU doctor or clinic you can contact when you're overseas in case you need assistance or run into problems
- ask your 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 to help with translation
- if you're travelling on an organised tour, ask your dietitian for a letter explaining your diet – the travel company may be able to have it translated 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, you may want to search the web or ask your clinic for information about PKU associations in the country you intend to visit.

Have you packed?

- ☐ formula or supplement
- ☐ mixing container
- ☐ scoop, tablespoon measures, if used
- ☐ vitamin and mineral supplements if needed
- ☐ prescription
- ☐ low protein foods
- ☐ customs letter
- ☐ scales, if needed
- ☐ diet information
- ☐ food for your trip

Chapter 16

Financial assistance

- PKU products
- government allowances

Financial assistance

PKU products

Amino acid supplements for people with PKU are available on prescription and are fully subsidised in New Zealand and Australia.

New Zealand

In New Zealand, some special foods for people with PKU are also available on prescription. Other foods can be purchased directly from the supplier (ask your dietitian for contact details/order forms). Foods available on prescription include Loprofin and other low protein products, e.g:

- Loprofin All Purpose Baking Mix
- Loprofin pasta spirals, spaghetti and macaroni
- Loprofin rice.

Australia

The following low protein products for people with PKU can be purchased directly from the supplier. Contact details/order forms for these products are available from your metabolic dietitian or nurse.

- Loprofin Breakfast Cereal Loops
- Loprofin Egg White Replacer
- Loprofin low protein breads
- Loprofin milk replacements
- Loprofin pastas
- Loprofin All Purpose Baking Mix
- Loprofin biscuits – savoury and sweet
- Agluten riso
- Aproten pasta varieties (annelini, rigatini, spaghetti, tagliatelle and lasagne)
- Aproten low protein cracker toast
- Aproten crispbreads
- Country Harvest Egg-like egg replacer
- Low Protein Cheddar Cheese
- Low Protein Mozarella Cheese
- Orgran low protein pasta varieties (rigatini, corn pasta, rice pasta)
- Pinnacle low phenylalanine bread mix

Financial assistance

Government allowances

New Zealand

Ask your doctor or social worker for advice on financial support that you may be eligible for, or contact Work and Income New Zealand (phone 0800 933 922, or visit www.workandincome.govt.nz).
The usual prescription charges apply when obtaining PKU amino acid supplements and low protein foods.

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.

1. **The Health Care Card**
Children with PKU are eligible for the Health Care Card which means they are entitled to prescription medicines at the lower (pensioner) rate. This will allow you to get the special PKU formulas on the lower prescription rate. (You will only pay for the prescription, not for the formula.)

2. **The Carer's Allowance**
Parents of children with PKU automatically receive the Carer's Allowance, which is paid through Centrelink. Income and assets tests are not applied, but parents must be Australian residents. The current rate is $92.40 per fortnight (July 2005).

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

3. **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 and Aged Care. This allowance, for individuals with an inborn error of metabolism (IEM), is to help offset their food costs. To qualify for the allowance the person must be on an appropriate diet for their PKU, submitting regular blood tests and attending clinic. The IEM Allowance is currently paid at the rate of $200 per month (July 2005) to people of all ages and is normally claimed once a child reaches six months of age. This allowance is also exempt from a means or assets test. Claim forms are available through the PKU clinic and need to be signed by your doctor.

Chapter 17

PKU Resources

- PKU Associations in New Zealand and Australia
- breastfeeding support and information
- overseas PKU associations
- other resources

Chapter 17

PKU Resources

PKU associations in New Zealand and Australia

PKU NZ

PKU NZ is a charitable trust set up to support those with phenylketonuria and their families. As well as a regular newsletter, PKU NZ organises social events, picnics and Christmas parties.
Contact: PKU NZ, P. O. Box 872, Auckland.

Australian associations

MDDA Australia (Metabolic Dietary Disorders Association)

MDDA is a national association for people with a variety of metabolic disorders including PKU. It provides support, social activities, conferences and a quarterly newsletter. Head office is in Victoria.

Contact: MDDA, P.O. Box 33, Montrose, Victoria 3765
Tel: 1800 288 460
Email: mddaaustralia@iprimus.com.au
Website: www.mdda-australia.org

The PKU Association of NSW

Based in 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: Current contact details and previous copies of their newsletter are available from the PKU Association of NSW, C/o Treasurer, 58 Aberdeen Road, St Andrews NSW 2566
www.pkunsw.org.au

Breastfeeding support and information

New Zealand

Practical breast feeding support is available from your local midwife or Plunket Nurse.

Plunketline offers free telephone support 24 hours day, 7 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. Books and leaflets are available and the organisation has a free lending library.

Contact: Phone (04) 471 0690 for the name of your local support group leader (and for telephone advice) or email lllnz@clear.net.nz
Visit the website www.lalecheleague.org for more information.

Australia

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

Australian Breastfeeding Association

Telephone counselling is available seven days a week via the breastfeeding helpline in all capital cities and some rural areas (phone numbers below). You can also look under Australian Breastfeeding Association in your local White Pages. (Some books may still have it listed as Nursing Mothers' Association of Australia).

ACT/Southern New South Wales	(02) 6258 8928
New South Wales	(02) 8853 4999
Queensland	(07) 3844 8977 or (07) 3844 8166
Townsville	(07) 4723 5566
South Australia and NT	(08) 8411 0050
Northern Territory counsellor contact line	(08) 8411 0301
Tasmania	(03) 6223 2609
Tasmania – North	(03) 6331 2799
Victoria	(03) 9885 0653

Visit the website, www.breastfeeding.asn.au, for information about all aspects of breastfeeding.

Overseas Associations

UK PKU association:

www.nspku.org

American PKU Associations:

www.pkunews.org
www.pku-allieddisorders.org
www.pkuil.org
www.pkunetwork.org
www.pkulatetx.com for late-diagnosed people with PKU

Other Resources

Washington PKU clinic:

http://depts.washington.edu/pku/resources.html

Texas PKU clinic:

www.dshs.state.tx.us/newborn/pku.shtm

Chapter 18

Sample letters

- for childcare, preschool and school teachers
- for parents of your child's friends and classmates
- for doctors, maternal and child health nurses, and Plunket nurses
- customs declaration

Chapter 18

Sample letters

These sample letters can be photocopied and given to teachers, parents of children in your child's class, carers and health professionals who need to know about your child's PKU. You can also use them as a basis for composing your own letters.

When you're travelling overseas, taking a letter from your doctor for customs officials will save time and confusion.

Sample letters

Dear Teacher

_____ has an inherited condition, called PKU (phenylketonuria). This means that his/her body is unable to break down protein in the usual way. PKU is a rare, non-contagious condition, which, left untreated, can result in irreversible brain damage. Fortunately, PKU can be treated completely by a restricted diet. _____ has been on this diet from birth. Apart from needing a restricted diet he/she is exactly the same as other children in the class. He/she is no more likely to suffer illness than any other child, and can participate in normal school/preschool activities.

PKU is different from an allergy. If a child with PKU consumes food he or she shouldn't eat, there is no immediate reaction, but there will be a build-up of debilitating effects over time. The treatment for PKU is a low protein diet plus a protein supplement. Exact proportions of protein are measured each day. In order for us to do this accurately, we would ask for assistance with the following, and for supervision at mealtimes:
** food should not be shared with other children*
** only food provided by us should be given (or foods from the list of foods that we have authorised as OK)*
** leftover food needs to be kept and brought home in the lunch box to help us in our daily calculations of the diet*
** if the protein supplement is taken to preschool/school it should be treated as a medicine, and we would like to discuss putting a system in place to make sure it is taken.*

If _____ has eaten any food that has not been provided by us it is important we are informed that day so we can make adjustments to the diet for the rest of the day.

Many thanks for your assistance. It is greatly appreciated. Please contact us on _____ if you need more information, or you can contact the PKU clinic on _____.

Kind regards

Sample letters

Dear Parents

_____ is a classmate of your child. She/he has a rare condition called PKU (phenylketonuria), which means her/his body cannot break down the protein from food in the usual way. PKU is an inherited condition she/he has had from birth. It is neither infectious nor contagious. You have probably never met anyone with PKU, although all babies are tested for it at birth.
Children with PKU are very normal and no different from any other child. The only way you would know they have PKU is from the type of food they eat.

_____ won't be eating many of the foods your child eats and it may even seem she/he eats unhealthily. Each day we carefully measure and keep track of the protein in the food she/he eats. Children (and adults) with PKU can't eat red meat, fish, chicken, eggs, milk, beans or nuts. They eat a lot of fruit and vegetables, special prescription bread and pasta, and they take a protein supplement every day to make up for what they don't get from the foods other people eat.

It is essential that they follow their diet carefully. When a child with diabetes or a food allergy eats the wrong food there is an immediate reaction. However, if a child with PKU eats the wrong food the problems happen over a long period of time.

We are used to providing most of what she/he eats wherever she/he goes. If you would like her/him to come and play or go to a party, we'll be happy to talk to you about what food to give her/him. We also know how to feed a child without PKU!

Kind regards

Phone:

Sample letters

Dear Doctor/Maternal and Child and Health Nurse/Plunket Nurse

Re: _____

This infant has been diagnosed with phenylketonuria (PKU) following a positive newborn screening test. PKU is a recessively inherited condition, characterised by a deficiency of the liver enzyme, phenylalanine hydroxylase, used to process the amino acid phenylalanine in protein foods.

Without treatment there is progressive, irreversible brain damage. When treatment commences soon after birth, normal intellectual and behavioural development can be expected.

Treatment:

- A very low protein diet and a phenylalanine-free amino acid formula/supplement are given. Prescriptions are required for the supplement and parents need to be meticulous with the diet.
- On diagnosis babies are generally given phenylalanine-free formula alone for several days to reduce serum phenylalanine (Phe) levels. A Phe containing feed – either breastfeeds or standard infant formula – is then added (given after the formula usually), to prevent phenylalanine deficiency.
- The aim is to maintain the serum Phe level within the recommended treatment range, with weekly monitoring via blood tests which are processed by the Newborn Screening lab in each state (Australia), and in Auckland (New Zealand).

Management:

- The metabolic team at _____ manages this child's PKU and is happy to discuss or advise on relevant issues. Please call if you have any concerns or questions.
- Infants with PKU need to be weighed every week for the first few weeks, and then monthly to ensure adequate growth. Solid foods, introduced from around 6 months, are individually tailored to ensure nutritional adequacy, based on serum Phe levels and tolerance for dietary protein. Specialised protein-free cereal and grain products are generally required.
- Except for the need for a strict dietary regimen, children with well-controlled PKU should remain well and healthy, with normal growth and development. However, common childhood infections or poor feeding and weight loss may cause a temporary rise in phenylalanine levels, which may need management.
- All regular childhood immunisations are recommended.
- Management of common illnesses should be as for any other child, however, wherever possible, medications should be checked for artificial sweeteners which contain phenylalanine. These sweeteners may be labelled as Aspartame; brand names incude Nutrasweet, Canderal,

Neotame or Additives 950, 951, 961 and 962. If an alternative medication without the sweetener is available this should be given.

- As the diet to best manage PKU is a time consuming and expensive regimen, an application for the Commonwealth Government Inborn Errors of Protein Metabolism Dietary Allowance and Carers Allowance will be submitted.

Outcome of early-treated PKU is good and current policy is to recommend that the PKU diet continue for life. Parents receive regular ongoing education and review regarding PKU and its dietary treatment. Blood test results are collected by the metabolic team.

Yours sincerely

Doctor:
Address:
Date

Customs declaration

Dear Customs Officer,

RE: _____ name
_____ DOB
_____ address
_____ medical record no.

_____ has a rare metabolic condition known as phenylketonuria (PKU) that affects around one in every 15,000 people. Management of this condition involves strict adherence to a low protein diet and an oral supplement called _____ (only available on prescription). This supplement contains an amino acid mix to compensate for the lack of natural protein in their diet.
Because this supplement must be taken daily it is necessary that a supply is carried while travelling internationally. It may also be necessary for supplies to be received from Australia by international courier/mail. If there are any concerns regarding this medical supplement please make contact with me or your local medical authorities. _____ also requires the following (only available on prescription):_____
_____
_____
_____

Yours sincerely,

Dr

Title

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 phenylalanine and should be avoided by people with PKU. It may be listed on food labels as Nutrasweet, Canderal or additive 951.

BH4

BH4 is short for tetrahydrobiopterin. PKU is caused by a deficiency in activity of the enzyme, phenylalanine hydroxylase (PAH). For PAH to function properly, BH4 is needed. In some people with PKU, taking BH4 supplements 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.

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. Phenylalanine is an essential amino acid.

Exchange

An exchange system is used in New Zealand to count phenylalanine in the diet. One exchange = 50 mg of phenylalanine.

Fat

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

Folic acid

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

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 by some clinics to keep track of phenylalanine intake. In most foods, one gram of protein contains 50 mg of phenylalanine.

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.

Newborn screening

All newborn babies in Australia and New Zealand have a blood test within the first few days of birth to measure phenylalanine levels (among other things).

PAH

The abbreviation for phenylalanine hydroxylase.

Phe

The abbreviation for phenylalanine.

Phe-free 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 phenylalanine), plus vitamins, minerals and extra tyrosine. It is vital for people with PKU.

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 phenylalanine in foods in the usual way, so they must avoid foods containing protein (and therefore phenylalanine). The small amount of phenylalanine they need comes from carefully measured amounts of low protein foods. If PKU is not treated, phenylalanine builds up in the blood causing brain damage.

Phenylalanine level

This is the measurement of phenylalanine in the blood. A normal blood phenylalanine level

is around 70 µmol/L. In untreated PKU, levels may reach 3000 µmol/L.

Phenylalanine hydroxylase

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

Phenylketonuria

An inherited condition where the body lacks the enzyme phenylalanine hydroxylase needed to break down phenylalanine in foods. The condition is treated from soon after birth with a special diet. Doctors recommend that this diet be followed for life.

Plunket Nurse

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

PKU

See phenylketonuria

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, 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 phenylalanine in the blood.

PKU Clinic Team

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

Protein supplement

See supplement.

Tyrosine

Tyrosine is an amino acid that is partly obtained from phenylalanine. 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 phenylalanine to tyrosine. They obtain tyrosine from the supplement.

Supplement

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

Unit

A system of counting units is used by many clinics in Australia to measure the phenylalanine in the diet. One unit of phenylalanine is the amount of food that contains 15 mg of phenylalanine.

PKU Quiz

Use this quiz to test your knowledge about PKU. Have a go at answering the questions first, then look at the answers at the end of the quiz. People with PKU have provided many of these answers and ideas – some of which you may like to adopt for yourself.

Q 1 How long should someone with PKU stay on the low protein diet and Phe-free supplement?
A _____

Q 2 What makes your Phe levels go up?
A _____

Q 3 What makes your Phe levels go down?
A _____

Q 4 Does playing sport or exercising more bring your levels down?
A _____

Q 5 How do you manage PKU when you're out? Do you plan ahead? Does it depend on who you're with or where you're going?
A _____
_____
_____

Q 6 What can you eat at your favourite café?
A _____

Q 7 What can you eat at takeaway outlets?
A _____

Q 8 What can you eat at your favourite restaurant? How would you manage if you were going to a restaurant that you hadn't been to before?
A _____
_____
_____
_____

Q 9 Your ideas for quick meals?
A _____

Q 10 Your ideas for barbecues?
A _____

Q 11 Your ideas for parties?
A_____
_____
_____
_____

Q 12 What do you do when you stay over at a friend's place? What do you need to think about?
A _____
_____

Q 13 How do you plan for camping trips?
A _____
_____

Q 14 What about weekends away? How would you manage if you were going away with people who didn't know about your diet or what you could eat?
A _____
_____

Q 15 What happens if you eat more Phe or protein than you are supposed to have when you are out or away for the weekend?
A _____
_____

Q 16 You have a girlfriend or boyfriend: what are you going to tell them about PKU? What are the important things for them to know?
A _____

_____

Answers

Q 1 How long should someone with PKU stay on the low protein diet and Phe-free supplement?

A In one word . . . **forever!**
It used to be thought that it was OK to stop the diet and supplement any time from six to ten years old. It was thought that by this time, high Phe levels would not cause any more damage to your brain or nervous system (the spinal cord and nerves). Newer research suggested that high Phe levels may affect adults' brain function and that staying on diet was beneficial. It is now recommended that people with PKU stay on a low protein diet and take the supplement **every day** for the **whole** of their life.

Stopping your diet and supplement and having high Phe levels may have side effects.
Some people with PKU who have gone off their diet and then start it again say that when they are back on diet they:
- feel better
- look better
- can think more clearly
- are less moody
- find it easier to get along with others (their friends and families say that they are easier to get on with as well!)
- feel less tired and have more energy
- can concentrate
- can think clearly to study and do exams
- can complete assignments more easily
- think it is a must for doing final year school exams
- can work things out better, e.g. when trying to think strategically such as in team sports or playing games or in jobs where you need to think clearly.
  Brain scans using magnetic resonance imaging (MRI) show changes in the brain when Phe levels in the blood are high. This improves when Phe levels are lowered. The effect of these changes in the long-term is not known as yet.

Some people with PKU who are not on diet develop problems such as:
- tremors (the shakes)
- nervous system problems such as behaving inappropriately or having

mood swings or being confused about reality – not seeing things as they really are
- stiff or weak legs
- headaches
- nutritional deficiencies, e.g. lack of vitamin B12, iron and calcium
- eczema.

If you stop your diet or supplement then it is essential – an absolute **must** – to talk to your doctor and dietitian so that you do it in the best possible way with the least amount of side effects and know what you are getting yourself into.

Eating a normal diet is very different from being on the PKU diet. It is unsafe just to avoid high protein foods. See the section on Safe Off Diet (chapter 11).

Q 2 What makes your Phe levels go too high?

A

- being sick
- dieting to lose weight
- not eating enough and losing weight as a result
- not having enough of your supplement
- eating too much protein from food

You'll find more information on blood Phe and what affects your levels in chapter 6.

Q 3 What makes your Phe levels go too low?

A Not having enough protein from food.

Q 4 Does playing sport or exercising more bring your levels down?

A Doing extra exercise, after eating too much protein, will not bring your Phe levels down. But exercise that builds muscle may increase the amount of Phe you can tolerate in food over time (see Exercise in chapter 11).

Q 5 How do you manage PKU when you're out? Do you plan ahead? Is it different depending on who you're with or where you are going?

A Eating is something we do every day and a lot of our socialising takes place around eating. Planning makes it easier to be prepared, so you can make sure you get enough of the foods you like that are also low in Phe or protein. Knowing what foods will be available and taking any supplies you might need can be helpful.

Another strategy is to eat a snack before you go out if you don't know what will be available. This saves you being hungry when there is nothing suitable for you to eat. It also means you can safely say you've already eaten and then just have a snack-type meal.

What makes a difference?
- who is present – new friends, old friends, people you want to impress
- how hungry you are
- how many questions you get from the people present
- being hassled to eat – some people think that because you're not eating much you have an eating disorder or are being rude.

Have you thought about what to tell your friends about PKU? What about new friends? Are you going to tell them only if you need to, or will you tell them anyway so it makes it easier to just be yourself?

Not letting on you have PKU could make managing your PKU more difficult.
What do you want to tell people about PKU? It works to say that you only eat low protein, mainly vegies and fruit, and that it is safe for you to do this – in fact important.
Is there anything else that you know works from experience?

Does it matter to other people that you have PKU?
Just in case you didn't know, having PKU is pretty amazing, not many people know about it and when people do find out, as you probably know already, they find it really interesting. Think about some of the questions that you get asked when people find out. Most of the time they can't imagine how you have managed all your life without eating meat and having to take a supplement every day.
If you've been generous enough to let your friends taste your supplement, you will know that most think you are even more amazing for being able to take it day in and day out all your life.

Remember, PKU is just one part of you. You have many qualities and PKU is not the only one that makes you unique.

Q 6 What can you eat at your favourite café?

A
- salad without red meat or chicken or fish or egg or cheese or tofu or nuts
- fruit
- fruit salad
- bread/roll – if you are able to have bread then a sandwich with salad or vegemite or banana or avocado, herb or garlic bread*
- bruschetta*
- chips*
- jacket potato* with low protein filling - tomato or guacamole or sour

cream and sweet chilli sauce
- vegetable soup (not based on milk, lentils or other beans, e.g. kidney or white beans or chick peas)
- pasta* with tomato sauce (no cheese – ask for no cheese or cheese separately)
- other pasta sauces that may be OK (check the description on the menu) are: napoletana (tomato with basil), arrabiata (with chilli), primavera (with vegetables)
- vegetable based risotto* – ask for no cheese
- side serve of vegetables

*These foods may be too high in protein if you usually have the low protein versions. If need be, rather than starving, have a small serve and be cautious about protein intake over the rest of the day.

Q 7 What can you eat at takeaway outlets?

A

- chips*
- apple pie*
- fruit salad
- salad
- soft drink (not diet)
- juices
- mashed potato* and gravy
- hash browns
- corn
- ice-cream*
- pizza* – ask for tomato and vegetables, no cheese; thin crust is lower in protein; many pizza places offer 'no cheese' pizzas, especially as there are more people with allergies these days.

Many takeaway food outlets label their food with the nutrient content so you can work out the protein content.

Commercial vegetable burgers are **not** low in protein.

*These foods may be too high in protein if you usually have the low protein versions. If need be, rather than starving, have a small serve and be cautious about protein intake over the rest of the day.

Q 8 What can you eat at your favourite restaurant? How would you manage if you were going to a restaurant that you hadn't been to before?

A

Australian/New Zealand restaurants

- vegetables or salad based meals, e.g. vegetable stacks, ratatouille
- vegetable based risotto*
- potato as chips, wedges or potato in jacket*
- pasta dishes with sauces based on vegetables, e.g. rocket, garlic, olives, artichokes, sweet potato, pumpkin and semi dried tomatoes – if you can take your own low protein pasta the chef may be able to cook that and serve it with one of these sauces*
- dressings or sauces, e.g. gravy or barbecue sauce
- bread/roll/salad wrap*
- fruit, e.g. caramelised figs, compote of fruit, strawberries, grapes in toffee

Italian restaurants

- pasta with tomato based sauces or sauces listed above or sauces with mushrooms and cream – no cheese
- garlic mushrooms
- vegetarian antipasto – artichokes, olives, tomatoes, eggplant, zucchini and capsicum
- stuffed vegetables* – check there's no meat or cheese
- fruit
- gelato based on water

Chinese restaurants

- boiled rice, plain fried rice, noodles*
- vegetable dishes, e.g. stir fried vegetables
- prawn crackers*
- lychees
- fresh fruit

Thai restaurants

- boiled rice, plain fried rice, noodles*
- vegetable dishes, e.g. green or red curry (avoid dishes with nuts, e.g. Panang curry)
- fresh fruit

Indian restaurants

- boiled rice, naan bread, chapatis, papadums, parathas*
- dishes made with sago
- vegetable dishes without legumes (lentils, red or white beans, chick peas)
- mango chutney and pickle, lime pickle

Malaysian restaurants
- boiled rice, plain fried rice
- vegetable dishes, e.g. stir fried vegetables, avoid vegetable dishes cooked with blachan (fish paste)
- sago based desserts, e.g. gula melaka
- fresh fruit

Greek restaurants
- rice, cracked wheat (bulgur), bread*
- vine leaves*
- dips – eggplant
- olives
- vegetable dishes, e.g. vegetable kebab
- salad

Mexican restaurants
- potato wedges*
- tortilla chips*
- corn chips,* taco shells*
- sour cream, salsa
- guacamole

*These foods may be too high in protein if you usually have the low protein versions. If need be, rather than starving, have a small serve and be cautious about protein intake over the rest of the day.
If you eat regularly at a restaurant or know ahead of time where you are going, you'll often find the restaurant will be happy to cook low protein food.

Q 9 Your ideas for quick meals

A (See chapter 10, Food on the Run)

Q 10 Your ideas for barbecues

A

- homemade vegetable and fruit skewers
- hash browns
- commercial vegetable fingers
- homemade vegetable patties
- homemade vegetable sausages
- corn on the cob in foil
- mushrooms
- eggplant slices
- dips, e.g. guacamole, salsa, eggplant
- take low protein crackers with you
- salads

Q 11 Your ideas for parties?

A Try to avoid high protein foods, fill up on the lower protein nibbles such as salad platters, tomato or avocado based dips. Offer to bring something along for everyone to share. Most people like vege chips, low protein dips and vegetable sticks. (See chapter 10 for more party food ideas.)

Q 12 What do you do when you stay over at a friend's place? What do you need to think about?

A Planning makes it easier. Tell them what you can and can't eat if possible. That way they don't prepare a huge meal which you then can't eat – a situation where everyone feels bad.

Take your supplement. Ready–to–go versions, such as tetrapaks or sachets that are easy to mix are the easiest, or take your supplement already measured out and mix it at your friend's place. If you can, you might want to take it before you go. If you normally have your supplement in the morning then delay this until you get home the next day and catch up what you missed out on.

Ask what is being served and take some extra food that you can eat if you think you'll need it. You don't want to be hungry. You might want to take extra to share. To make sure you get enough to eat, take along a bag of vege chips, some fruit (fresh fruit and individual fruit snack packs), PKU bread/buns, snack bars and low protein biscuits and crackers.

Q 13 What about camping trips?

A Supplement:

Take enough of your supplement for the number of days you will be away and a bit to spare. Supplement provides energy – essential if you are busy outdoors.
Supplements that are in sachets or ready-to-drink or that you take as capsules that don't need refrigerating are handy. If you need to make up your drink from powder it is a good idea to measure it out in individual portions in airtight and waterproof containers, such as sandwich bags or plastic containers, before you go away. If you're planning to take your supplement twice a day, then measure it into two portions for each day. You will need to take enough water to mix with your powder.
Food:
Plan what food you will take. Plenty of low protein food will help you keep your energy levels up.

Here are some ideas:
- dried fruit
- individual fruit snack packs

- fresh fruit
- low protein noodles
- low protein pasta
- ready made pasta sauces
- low protein cereal – eat it dry
- rice milk
- snack bars
- low protein biscuits and crackers
- vege chips – pack in a plastic container so that they don't get crushed
- cherry tomatoes
- rice cakes or corn thins
- lollies (without milk or chocolate)

Q 14 What about weekends away? How would you manage if you were going away with people who didn't know about your diet or what you could eat?

A Planning makes it easier. Take your supplement. Ready–to–go versions such as tetrapaks or sachets that are easy to mix are the easiest, or take your supplement already measured out and mix it at your destination.

Food:
Talk to whoever is organising the food. Ensure that there will be food that you can eat. If not then you will need to take extra food just for you, or you might want to take extra to share. You don't want to be hungry. See the answers to questions 12 and 13 for more tips and ideas.

Q 15 What happens if you eat more Phe or protein than you are supposed to have when you are out or away for the weekend?

A If you are over in your protein intake for one day, you can cut back the next day. Only go over by 10 % or one tenth more than your usual protein if you can manage it. This means that if you are used to having 25 units, 10 % = $^{10}/_{100}$ x 25 = 2.5 units. Similarly, if you count grams of protein or exchanges 10 % of 20 g = $^{10}/_{100}$ x 20 = 2 g protein or 2 exchanges.

Q 16 You have a girlfriend or boyfriend. What are you going to tell them about PKU? What are the important things for them to know?

A It is essential that they know about PKU, especially if you are spending a lot of time with each other.

Important things to tell are:
- what PKU is
- what you can eat
- about your supplement

Offer them your handbook to look at.


You can never have *too many choices.*
SHS

Less volume, More choice.

Introducing our: **LOW-VOLUME RANGE**

PK MAX

A low-volume choice for children.

For children 1-10 yrs.

Unflavoured, Tropical or Raspberry flavour.

1 sachet + 40mL water makes a semi-solid dessert or can be taken as a low-volume drink by adding more water.

Presented in convenient sachets.

Should be consumed with water or diluted drinks.

LOPHLEX

Low in volume, low in calories.

For children over 8 years, adolescents and adults including pregnant women with PKU.

Unflavoured, Orange or Berry flavour.

1 sachet + 65mL water, means only 80mL to drink. Best served chilled.

Presented in convenient sachets.

Should be consumed with water or diluted drinks.

NUTRICIA

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498

Amino Acid Supplements SHS Loprofin

PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET

NC113a - JULY 05

Ready to use, ready to go.

CHOOSE CONVENIENCE

EASIPHEN

Portable, convenient,
best served chilled.

For children over 8 years, adolescents and adults, including pregnant women with PKU.

Forrest Berries or Grapefruit flavour.

Presented in 250mL pre-mixed tetra pack with straw.

Can be easily stored in the cupboard at home. Refrigerate after opening.

NUTRICIA

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498

Amino Acid SUPPLEMENTS SHS Loprofin

PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET

Tried & Trusted

LOTS OF OLD FAVOURITES TO CHOOSE FROM

XP ANALOG

Birth - 1 year.
Unflavoured.

XP ANALOG LCP

Contains LCPs
(Omega 3 and Omega 6 fats).
Birth - 1 year. Unflavoured.

XP MAXAMAID

For children 1 - 8 years.
Unflavoured or Orange flavour.

MINAPHLEX

Includes a balanced blend of essential fatty acids.
For children 1-10 years.
Unflavoured or Pine-Vanilla flavour.
1 sachet + 100mL water. Presented in convenient sachets.
Best served chilled.

XP MAXAMUM

Choose tin or sachets.
For children over 8 years and adults, including pregnant women with PKU.
Unflavoured or Orange flavour.

NUTRICIA

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498

Amino Acid SUPPLEMENTS SHS Loprofin
PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET

NC113b - JULY 05

Phlexy-10 mix 'n' match

DRINK IT, TAKE IT, EAT IT, YOU CHOOSE

The Phlexy-10 system is a flexible dose-related interchangeable range of products for Phenylketonuria. This system has incorporated the need for choice, convenience and low volume in addition to providing the amino acids in a more palatable form.

For children and adults. Products require additional source of vitamins, minerals and trace elements. Intake should be determined by clinician or dietitian only. Should be consumed with water or diluted drinks.

PHLEXY-10 DRINK MIX

Apple & Blackcurrant or Tropical Surprise flavour.

1 sachet + 100mL water.

Powdered drink mix in convenient sachets.

PHLEXY-10 BARS

Citrus fruit flavour.

PHLEXY-10 CAPSULES

PHLEXY-10 TABLETS

Unflavoured

NUTRICIA

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498

Amino Acid SUPPLEMENTS SHS Loprofin
PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET

LOPROFIN

A huge *choice* of low protein, high-enjoyment foods.

Nutricia Australia is proud to bring you the most comprehensive range of low protein foods available, to help you manage your low protein diet.

BISCUITS

Individually wrapped portion packs – keeping them fresh for longer.

PASTA

Authentic Italian low protein pasta. Delicious with tomato-based sauces.

CHEESES

Great sliced with your favourite Loprofin crackers.

BREAD

Sliced for toast or sandwiches, or part baked bread rolls, delicious straight from the oven for that home-baked taste.

NUTRICIA

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498

Amino Acid SUPPLEMENTS SHS Loprofin

PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET

NC113c - JULY 05

A huge *choice* of low protein, high-enjoyment foods.

DELIVERED STRAIGHT TO YOUR DOOR.

Check with your dietitian about the suitability of new foods before introducing them into your diet.

MILK REPLACEMENTS

Use it on cereal, in milkshakes, soups and sauces or even in baked goods.

BAKING NEEDS

Great for all your baking needs, including cakes, biscuits and meringues.

CONVENIENCE FOODS

Sweet and savoury snacks for when you are on the go!

BREAKFAST CEREAL LOOPS

Great on its own as a snack or with low-protein milk and your favourite fruits for a quick and tasty breakfast.

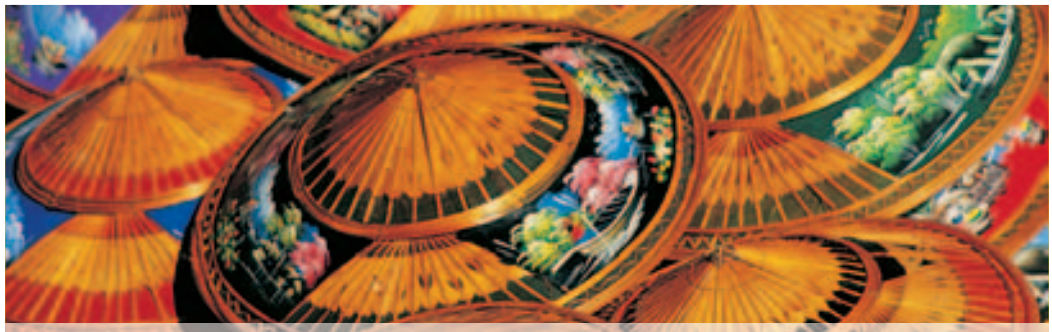
Choose from our *huge range of foods.*

NUTRICIA

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498

Amino Acid SUPPLEMENTS SHS Loprofin

PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET



YOUR CHOICE

Nutricia Australia is proud to bring you the most comprehensive range of amino-acid supplements and low protein foods, to help you manage your low protein diet.

Nutricia's amino acid supplements are available only on prescription.

For samples contact your PKU clinic. For supplies, your pharmacist should contact their usual wholesaler or Nutricia Customer Service on: ph: 02 8853 9696 fax: 02 9894 6498.

The Loprofin range of products can be ordered directly from Nutricia and delivered straight to your door.

Additional services / materials available from Nutricia include:

- o Living with Loprofin newsletter.
- o Product catalogue and Order Forms.
- o Low Protein Collection Cookbook - a ring bound folder designed to receive additional recipe inserts.
- o Bread Making Made Simple booklets.
- o Clinical Care Line – 5 days a week, freecall service to answer any questions you may have about the Nutricia range of products **1800 060 051.**

NUTRICIA

Amino Acid SUPPLEMENTS SHS Loprofin

PROVIDING MORE CHOICE FOR THE LOW PROTEIN DIET

NC113d - JULY 05

A Guide to using

PKU express

A Phenylalanine free protein substitute for use in the dietary management of Phenylketonuria. For people aged 8 years and above.

Packaged in re-sealable foil pouches. Available in two options: OrangeCooler and PurpleCooler.

Packaged in individual sachets. Available in Blackcurrant, Lemon, Orange, Raspberry, Tropical and Unflavoured.

Contains all the necessary vitamins and minerals to meet the needs of this age group. Each pouch and sachet is equivalent to 15g protein making it easy to swop one for the other.

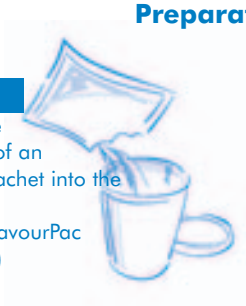
No weighing! Low volume! No fuss!

Simply Shake PKU express™ RANGE

Preparation guidelines for express powder

STEP 1

Empty the contents of an express sachet into the beaker.
(plus 1 FlavourPac if desired)



STEP 2

Fill the beaker about ½ way up with cold water, approximately 80ml. Secure the lid and shake well.

STEP 3

Drink immediately.

For best results the product should be freshly prepared and when possible used immediately.
However if necessary store in a refrigerator and use within 24 hours - re-shake before use.

FLAVOUR IDEAS

To add more variety here are a few suggestions.

The unflavoured express can be flavoured using the Vitaflo FlavourPac sachets.
Available in blackcurrant, lemon, orange, raspberry or tropical.

Simply add 1 flavour sachet to the unflavoured express and make up as illustrated.

To give a stronger fruitier flavour, permitted cordial or fruit juice can be added to the already flavoured express giving you a choice of taste.

Preparation guidelines for express liquid

Shake, Open, Drink.
Refrigerate if desired.
PKU express liquid can also be frozen overnight and will take approximately 2½ hours to defrost.

IMPORTANT - WATER OR PERMITTED DRINKS SHOULD BE TAKEN AFTER THE EXPRESS.

Vitaflo

Innovation in Nutrition

AUSTRALIA · CANADA · FRANCE · GERMANY
IRELAND · ITALY · SCANDINAVIA · SPAIN · UK · USA

Manufactured in the UK for Vitaflo International Ltd.
Vitaflo Australia PTY LTD
PO Box 8067, Newtown, Victoria 3220

Tel: 03 5244 5811 Fax: 03 5244 5822

E-mail: vitaflo@vitaflo.com.au Web: www.vitaflo.co.uk

Code: 0605

A Guide to using PKU gel

A phenylalanine free gel or drink for use in the dietary management of Phenylketonuria.
For children of 12 months to 10 years of age.

Packaged in individual sachets, it is convenient and simple to prepare.

Simply shake the solution in the beaker provided. This makes it easy for you or your child to prepare anywhere - childcare, school, days out, and holidays.

PKU gel can be taken as a smooth gel or a low volume drink whichever suits your child.

Available in:
Blackcurrant, Lemon, Orange, Raspberry, Tropical or Unflavoured Sachets.

Contains all the necessary vitamins and minerals to meet the needs of this age group.

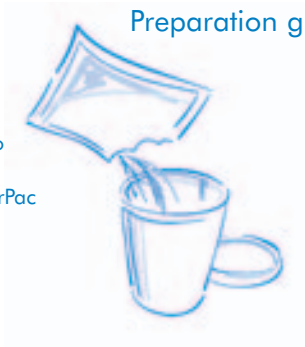
Each sachet of powder is equivalent to 8.4g protein.

No weighing!
Low volume!
No fuss!

Simply Shake gel™

Can be taken as a gel or a low volume drink, whichever suits you.

Preparation guidelines for gel



STEP 1

Empty the contents of a gel sachet into the beaker.
(plus ½ - 1 FlavourPac if desired).

STEP 2

FOR GEL

Pour 30ml of cold water into the beaker.

FOR DRINK

Fill the beaker ½ way up with cold water, approximately 100ml.

STEP 3

Secure lid and shake the beaker well.

STEP 4

FOR GEL

Leave to stand for 2 minutes for the gel to form.

FOR DRINK

Drink immediately.

FLAVOUR IDEAS

To add more variety here are a few suggestions.

The unflavoured gel can be flavoured using the Vitaflo FlavourPac sachets. Available in blackcurrant, lemon, orange, raspberry or tropical.

Simply add ½ - 1 flavour sachet to the unflavoured gel and make up as illustrated.

To give a stronger fruitier flavour, permitted cordial or fruit juice can be added to the already flavoured gel giving you a choice of taste.

WEANING GUIDE

PKU gel is a great low volume supplement to introduce to your baby around 1 year of age either as a gel from a spoon or in low volumes from a cup. It allows your baby to develop cup feeding skills and move away from dependence on bottles. The small volume allows your baby to develop hunger and learn to eat allowed foods for meals and snacks, avoiding her filling up on large volumes of liquid. If preferred the supplement can be given as a gel from a spoon as a snack or part of a meal. PKU gel is ideal for toddlers and young children too due to its low volume and versatility. Not filling up on large volumes allows them to develop normal eating habits and behaviours.

For best results the product should be freshly prepared and when possible used immediately. However if necessary store in a refrigerator and use within 24 hours - re-shake before use.

IMPORTANT - WATER OR PERMITTED DRINK SHOULD BE TAKEN AFTER THE GEL OR DRINK.

Vitaflo

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E-mail: vitaflo@vitaflo.com.au Web: www.vitaflo.co.uk

Code: 0605

Phenex - 1®

Phenex - 2®

Formulas for nutrition support for infants, children and adults with PKU

Features and Benefits;

- Amino acid modified formula for infants and toddlers with PKU
- Unflavoured powder
- Comes in 350g can
- PBS listed

Features and Benefits;

- Amino acid modified formula for children & adults
- Delicious vanilla flavour or unflavour powder options
- Comes in 350g can
- PBS listed

For help with supply to your chemist please call

NSW/QLD - Brigitte Rael on 0407 101 580
VIC/SA - Martene Harvey on 0409 457 589
WA - Jacqui Thomas on 0409 045 228

Abbott Australasia Pty Ltd.
ABN 95 000 180 389
32 - 34 Lord Street, Botany NSW 2019 Austral
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Customer Service 1800 225 311

A Vision for Better Nutrition

ABBOTT

PKU contacts

Doctor

Name:

Contact details:

Address:

Name:

Contact details:

Address:

Name:

Contact details:

Address:

Dietitian

Name:

Contact details:

Address:

Name:

Contact details:

Address:

Name:

Contact details:

Address:

Nurse

Name:

Contact details:

Address:

Name:

Contact details:

Address:

Blood laboratory/Newborn Screening

Name:

Contact details:

Address:

Social worker

Name:

Contact details:

Address:

Family doctor

Name:

Contact details:

Address:

Chemist

Name:

Contact details:

Address:

PKU support groups

Name:

Contact details:

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Contact details:

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PKU Handbook

Name:

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