The ASIEM
Low Protein Handbook for Tyrosinaemia

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Introduction

This handbook contains information relevant to the management of many disorders affecting the metabolism of protein.

Some of the sections are specific to the condition that you or your child has. Many sections cover a range of conditions that are managed in the same way—in these sections the condition is called ***metabolic disorder or metabolic condition***.

The common sections are relevant for the following metabolic conditions:

- Urea cycle disorders
  - Carbamyl phosphate synthetase (CPS) deficiency
  - N-acetylglutamate synthetase (NAGS) deficiency
  - Ornithine transcarbamylase (OTC) deficiency
  - Arginosuccinicaciduria
  - Arginase deficiency
  - Arginosuccinic acid synthetase (Citrullinaemia type 1)
- Maple syrup urine disease (MSUD)
- Tyrosinaemia type 1 and 2
- Homocystinuria
- Organic acidaemias
  - Glutaric aciduria type 1
  - Isovaleric acidaemia
  - Methylmalonic acidaemia
  - Propionic acidaemia
  - Holocarboxylase synthetase deficiency
- Hyperornithinaemia (gyrate atrophy)

Information in this handbook should only be used in conjunction with and should not replace advice from your Metabolic Team.

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- ☐ Chapter 2 How the diagnosis will affect my baby and our family
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Chapter 1

What is tyrosinaemia?

Finding out about tyrosinaemia

Tyrosinaemia is a treatable condition. People with tyrosinaemia cannot break down protein in the usual way. When tyrosinaemia is detected early and treated, children are able to remain in good health.

This chapter covers two types of tyrosinaemia – type 1 and type 2. These disorders have very different effects in the body. There are some other much rarer forms of tyrosinaemia (type 3 and Alkaptonuria) which are rarely seen in children.

Your child has: Tyrosinaemia Type……………..

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

This handbook explains the important things about tyrosinaemia, how it can be managed, and the support and help available to you. It also includes information, advice and encouragement from people who live with tyrosinaemia and other disorders that affect protein metabolism.

Managing tyrosinaemia is about keeping your child healthy. Once you are used to the management of tyrosinaemia and the diet, it is less overwhelming and daunting, and, as a parent, you can get on with raising and enjoying your child.

What causes tyrosinaemia

Tyrosinaemia is an inherited disorder, caused by a faulty gene. It is very rare. Only one or two babies a year are diagnosed with tyrosinaemia in Australia and New Zealand. For a child to inherit tyrosinaemia both parents must carry the faulty gene; boys and girls have an equal chance. (Tyrosinaemia inheritance is further explained at the end of this chapter. The glossary at the end of this handbook gives simple explanations of medical terms).

People with tyrosinaemia are born with a deficiency of one of the enzymes involved in the breakdown of tyrosine in the body. **Tyrosine** is an amino acid, found in foods containing **protein**.

Tyrosinaemia type 1 and **tyrosinaemia type 2** are caused by deficiency of different enzymes:

Tyrosinaemia type 1 is caused by deficiency of the enzyme **fumarylacetoacetase** (sometimes called fumarylacetoacetate hydrolase) and, **if untreated,** damages the liver and kidneys and brain which can result in liver failure or cancer and death during childhood.

Tyrosinaemia type 2 (sometimes called Richner-Hanhart syndrome) is caused by deficiency of the enzyme **tyrosine aminotransferase**. **If untreated** crystals of tyrosine build up in the eyes causing painful inflammation. High tyrosine levels also cause a build up of skin tissue (callouses) on the palms of the hands and soles of the feet. There have also been some reports in the medical literature of slow intellectual development in some children, but this has not been the case in any patients seen in Australia.

High tyrosine levels cause sore eyes and callouses on the skin

Not only are the two types of tyrosinaemia very different, but the severity of symptoms of the same type of tyrosinaemia can vary even between affected members of the same family.

Tyrosine is an amino acid, found in foods containing **protein**. Protein is made up of around twenty separate building blocks called amino acids. High protein foods include dairy products, meat, chicken, fish, eggs, soya products, nuts, beans and lentils.

Usually, when we eat protein, it is broken down into different amino acids which are used for growth, functioning and repair of the body. Some amino acids are broken down further, particularly if we eat more protein than we need or when we are sick.

Because people with tyrosinaemia can’t process tyrosine adequately, levels of this amino acid and other harmful metabolites (breakdown products) build up in the blood and damage the body.

Tyrosine is also made in the body from another amino acid called **phenylalanine** so too much phenylalanine is also a problem.

Phenylalanine as well as being part of protein in food is also found in some artificial sweeteners used in foods and medicines. See Chapter 3 for information on avoiding this source of phenylalanine.

People with tyrosinaemia need *some* tyrosine and phenylalanine, but only a small amount.

The science behind the condition and its management is explained in more detail at the end of this chapter.

How tyrosinaemia is detected

All babies born in Australia and New Zealand are tested for tyrosinaemia soon after birth, usually on the second or third day, as part of the newborn screening heel prick test. If the results are high, more tests are done to confirm that the baby has tyrosinaemia and which type.

Type 2 is easily detected by these tests but at the moment it is not possible to guarantee that all cases of tyrosinaemia type 1 will be detected. Testing for tyrosinaemia has only recently been introduced in Australia and New Zealand, so children born before the start of screening will not have been tested at birth.

Diagnosis in other babies, children or adults occurs when symptoms suggest to the doctor that the person has tyrosinaemia. Some newborn babies with severe forms of tyrosinaemia type 1 can become very sick, very quickly.

As soon as the diagnosis is made, treatment will be started. The baby will be given a special formula, also called a supplement, to lower the tyrosine to a safe level, and medications if needed.

When the disorder is detected with newborn screening, your baby has only been exposed to high levels for a short length of time. Before birth the mother's blood circulation can deal with the baby's tyrosine and the level stays within the normal range. Immediately after birth it starts to rise.

When tyrosinaemia is detected early and treated, children are able to remain in good health.

The treatment for tyrosinaemia

Tyrosinaemia Type 1

Tyrosinaemia type 1 is treated with a medication called NTBC and a special diet.

NTBC blocks an enzyme higher up in the tyrosine breakdown chain than the block in tyrosinaemia type 1. This stops the build up of harmful metabolites that damage the liver, kidneys and brain.

NTBC does not lower tyrosine levels so a diet low in tyrosine and phenylalanine is needed to prevent the potential damage caused by high tyrosine levels.

NTBC, particularly if started early, greatly reduces the risk of early development of liver cancer – whether it does in the long term is not yet known. It seems to be a safe treatment with side effects not being described at the time of writing.

Before the introduction of NTBC a liver transplant was the only effective treatment for tyrosinaemia type 1, as diet alone improved the child’s health but did not prevent the liver cancer.

Tyrosinaemia Type 2

Tyrosinaemia type 2 can be successfully treated with a diet low in tyrosine and phenylalanine. NTBC is not effective in tyrosinaemia type 2 as the block in type 2 is before NTBC’s point of action.

‘Tyrosinaemia type 1 and 2: the science behind the conditions and management’ at the end of this chapter explains this in more detail.

The special diet for tyrosinaemia

The special diet for tyrosinaemia is a low protein diet and a special supplement, both of which need careful monitoring. Regular blood tests to measure tyrosine and phenylalanine levels and regular attendance at the Metabolic Clinic are part of the treatment.

For healthy growth we all need to eat carbohydrate, fat, protein, minerals and vitamins. People with tyrosinaemia can't break down the large amount of tyrosine in protein foods, so the amount of protein in the diet is **restricted**. A special supplement replaces some of the protein, minerals and vitamins that would normally be in the diet. The supplement contains all the amino acids except tyrosine and phenylalanine. Sometimes a supplement of phenylalanine is prescribed which can be added to the amino acid supplement. The amount of protein tolerated and other nutrients will come from food.

See the Five Food Groups on page 8, which shows how the low protein diet varies from a normal diet.

It is also important to avoid some artificial sweeteners which contain phenylalanine. These are used in some foods and medicines.

Babies diagnosed with tyrosinaemia soon after birth grow up accepting the taste of the special supplement. Children diagnosed later can be fussier and find it harder to take. The Metabolic dietitian will advise you on ways to give the supplement.

Feeding your baby

Babies with tyrosinaemia are fed with two types of milk:

- breast milk or a standard infant formula
- a special formula that contains all the amino acids except phenylalanine and tyrosine.

The amount of each will depend on your baby's appetite and blood test results. Your doctor and dietitian will decide – all babies are different. The precise amount of tyrosine and phenylalanine a child with tyrosinaemia can tolerate varies from child to child. It also varies as the child grows.

Feeding your toddler

When solid food is introduced at around six months, babies with tyrosinaemia need to eat mainly fruits and vegetables, as well as certain baby foods. Later on, they can eat a wider range of low-protein foods, which may include special bread and pasta, and other special low-protein products. The Metabolic dietitian will help you manage the diet.

The amount of protein your toddler can tolerate from food is worked out by measuring blood levels of tyrosine. The Metabolic doctor or dietitian will tell you how much to give and will alter the intake as needed. The special formula will be changed to a special supplement designed for the needs of young children and the dietitian will work out how much of this is needed.

It can be more difficult to monitor food intake with toddlers because they are on the go and can become fussy with food. The clinic and other parents can give you tips to manage this.

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

Growing up with tyrosinaemia

Your child’s treatment for tyrosinaemia needs to be continued for life, to prevent damage to the body.

In all other ways you will be able to treat your child exactly the same as other children. A child with tyrosinaemia will enjoy things all children enjoy. A child with tyrosinaemia, diagnosed and treated early, has the potential to grow up and excel in whichever area they choose.

The Metabolic Team – usually a doctor, dietitian, nurse, laboratory scientist and sometimes a social worker – provides ongoing support for you and your child. They will keep you up-to-date with any developments in treatment of these disorders.

The Five Food Groups

Five food groups	Low protein food guide
◆ fruit & vegetables	◆  fruit & vegetables 
◆ breads and cereals	◆  low protein and special bread, flour & pasta
◆ meat, chicken, fish	
◆ milk & dairy	 ◆ special supplement 
◆ fats & sugars	◆ fats & sugars 

Comparison of the Five Food Groups and the Low Protein Food Guide

Teenagers and adults with tyrosinaemia

Treatment for tyrosinaemia type 1 and type 2 is for life. There may be serious consequences if protein intake is increased or the supplements and medications stopped. Changes in treatment will be needed at different ages and stages so regular contact with the Metabolic Team is important.

Children need to gradually learn about and take responsibility for the treatment of their tyrosinaemia – see Chapter 11. It is essential that teenagers and adults understand their disorder and its management. The Metabolic Team understands that different ages have different needs and will work with you so that the treatment best fits into your lifestyle.

Other illnesses and immunisations

Children with tyrosinaemia get the same number of coughs, colds and childhood illnesses as other children. Illness may cause a temporary rise in the blood tyrosine levels, but this does not cause long term harm. Tyrosinaemia is **not** one of the metabolic conditions with which children can get very sick with the usual childhood illnesses or accidents. Most medicines (preferably those that do not contain phenylalanine) can be given safely. Check with your Metabolic doctor if you are uncertain. For more information on managing illness see Chapter 3.

Routine immunisations should be given at the usual times.

Making sense of blood test results

Keeping the tyrosine and phenylalanine at just the right level is a balancing act. If the level is OK *most* of the time, your child’s health will not be affected by occasional low or high levels.

From time to time blood levels tyrosine and phenylalanine do vary. This may be because of illness, or changes in growth or food intake. Occasional higher or lower levels are unlikely to be a problem when overall control is good. Be guided by the Metabolic Team to keep to:

- Tyrosine levels ideally between 200-400 µmol/l and certainly below 600 µmol/l
- Phenylalanine levels in the normal range. Sometimes it is necessary to give supplements of phenylalanine to get the balance right

Other tests will be ordered by your doctor as needed. These include blood tests of liver and kidney function, liver scans and eye tests.

'It was so hard and scary at first finding out your child has this metabolic disorder and not knowing anything about it. Weighing, checking and writing down everything she eat and drank was very hard and time consuming in the first few years. But now we can tell how much protein is in most foods. We didn't think it would ever happen but our little girl is healthy and managing the disorder is easy now'

Genetics and tyrosinaemia—frequently asked questions

Will other family members be affected?

Other family members can be tested for the disorder and any further babies will be tested soon after birth. There is a one in four chance that a full brother or sister of a child with tyrosinaemia will also have the disorder. Testing during a further pregnancy (prenatal testing) may be possible. Discuss this with your Metabolic doctor or a genetic counsellor.

What is the role of genes?

Tyrosinaemia 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. Children born with tyrosinaemia have inherited a faulty gene from each parent.

What happens when genes are faulty?

Everyone carries some faulty genes, usually without knowing which ones. A person who has a faulty gene, but is not affected by it, is called a 'genetic carrier' or 'carrier'.

In tyrosinaemia, 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 with the same faulty gene have children, their child may be affected.

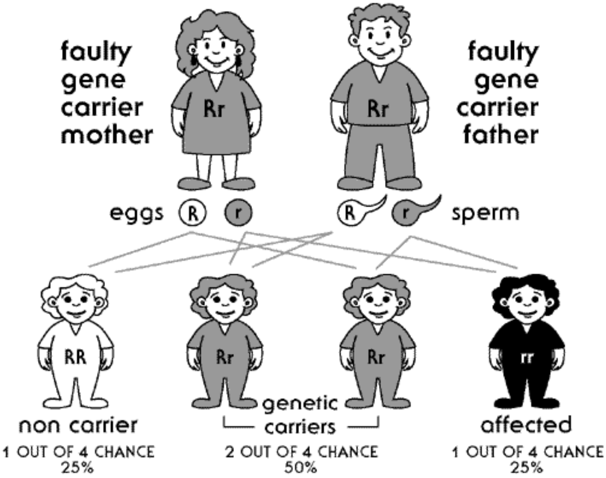
What faulty genes cause tyrosinaemia?

The faulty gene in tyrosinaemia controls one of the enzymes needed to breakdown tyrosine in the body. With two faulty tyrosinaemia genes in their cells and no regular ones, people with tyrosinaemia cannot make functioning enzyme in sufficient amounts. This affects the way the body deals with tyrosine.

What does having this faulty gene mean for the family?

The pattern of tyrosinaemia 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 healthy carrier like their parents
- a one-in-four chance of not carrying the faulty gene at all.



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

Reproduced with permission from Centre for Genetics Education Autosomal Recessive Inheritance Fact Sheet 8. Available from http://www.genetics.edu.au (Accessed September 2007).

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

If you have tyrosinaemia and your partner does not, the chance of having a baby with tyrosinaemia is very low. For this to happen your partner would have to be a carrier which is rare.

Testing for carrier status in a partner, from a family with no history of tyrosinaemia, is difficult and not always accurate. Prenatal testing is usually possible. Discuss testing with your Metabolic doctor or a genetic counsellor.

Tyrosinaemia type 1 and type 2: the science behind the conditions and management

The tyrosine and other harmful metabolites that build up in tyrosinaemia come from protein. All protein is composed of an assortment of around twenty chemicals called amino acids. One of these is *tyrosine*. We get tyrosine from the protein in our food and the body also makes it from another amino acid called *phenylalanine*.

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

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

In people who don’t have tyrosinaemia, some of the tyrosine they get from food and from phenylalanine is converted into energy in a series of steps using several different enzymes.

People with tyrosinaemia

People with tyrosinaemia cannot break tyrosine as one of these enzymes is missing.

The step at which this breakdown is blocked is different for tyrosinaemia type 1 and type 2. In both types build up of tyrosine caused by the block is toxic to the eyes and skin (pressure areas). The condition is called tyrosinaemia because when untreated there are high levels of tyrosine in the blood (- aemia means in the blood).

In tyrosinaema type 1 the enzyme that is missing is the last one in the breakdown chain so other metabolites (breakdown products) build up as well as tyrosine. These include maleylacetoacetate, fumarylacetoacetate as well as succinylacetone and succinylacetoacetate which are made from them. In high quantities these are very toxic compounds and damage the liver, kidney and brain.

Treatment of tyrosinaemia

The low protein diet and special formula/ supplement used in the treatment of type 1 and type 2 provide enough tyrosine and phenylalanine for growth and to keep the blood levels of tyrosine and phenylalanine in the safe range.

NTBC (full name is 2-(2-nitro-4-trifluoromethylbenzoyl)-1,-3-cyclohexanedione) is used in the treatment of type 1 but is not useful for type 2. It acts at an earlier step in the breakdown chain than the ‘block’ in tyrosinaemia type 1. This prevents the build up of the harmful metabolites maleylacetoacetate, fumarylacetoacetate, succinylacetone and succinylacetoacetate but does not lower tyrosine levels. A low protein diet and special formula/ supplement are still needed to lower tyrosine levels.

Chapter 2

Homocystinuria, tyrosinaemia & hyperornithinaemia: how a diagnosis of this disorder will affect your baby and your family

This information is for babies, children and adults with the following metabolic disorders:

- homocystinuria
- tyrosinaemia type 1 and type 2
- hyperornithinaemia

Your feelings

If your baby has been diagnosed by the newborn screening test, accepting the diagnosis can be difficult because your baby looks well, even perfect. It is alarming to be told that this disorder can interfere with your child's development and health. The words used are difficult to understand, and it may not be clear at first what the diagnosis really means.

As the good news that the disorder can be controlled starts to balance some of the initial distress, the amount of information about the treatment can be overwhelming.

If the disorder has been diagnosed because of symptoms of the disorder, you may be relieved that a diagnosis has been made. Even so the information can still seem overwhelming.

'I was really upset. The word 'devastating' comes to mind. 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 do something about this. After that we just went onward and upward. Sometimes I still feel sad for her but I don't let her see it.'

The first few days or weeks can be stressful. It is natural to feel worried about how you will manage. 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 metabolic disorder is a grieving process for many parents.

As you learn about the genetic basis of metabolic disorders, 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 metabolic disorder 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 metabolic disorder is not something you could have avoided, nor is it the result of anything you have done.

Taking one step at a time

Learning to manage your child's metabolic disorder helps in the process of coming to terms with the diagnosis. Most parents begin to feel more positive once they see their child's blood test results improve and start to see how the disorder is controlled.

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.

If your baby has been diagnosed as a newborn 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 disorder.

Sharing the experience

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

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

Seeing other children with the same disorder who are growing and developing well is reassuring and can help in developing confidence about the future. Your Metabolic Team can put you in touch with other families. Joining a parent group can be a support. It is important to remember that the symptoms and effects of metabolic disorder can be very different from child to child – even with the same disorder or from the same family. Sometimes reading about the disorder on the internet can be unnecessarily alarming. Talk to your doctor about the likely outcome for **your** child.

A team approach

A metabolic disorder 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 Metabolic Team is crucial.

A child's development and health is put at risk if the family is unable or unwilling to follow the advice of the Metabolic Team and the metabolic disorder is not well managed. Experience shows it's the children from families who seek medical help, attend clinic and have regular blood tests that do the best.

The metabolic disorder support circle: you will not be alone in managing your child's disorder.

Explaining the metabolic disorder

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

If your baby has been diagnosed soon after birth you may feel ready to tell family and friends about the diagnosis soon after it is made. But you may want to avoid telling too many people until you have adjusted to the diagnosis and the extra responsibility it brings. This allows you to let others know about the condition in your own time.

'I realised later that ringing everyone and telling them about the diagnosis and consoling them on the phone, made my own acceptance faster. The more people I told and said, look it's alright, we can cope, the more I was reassuring myself it was OK.'

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

If diagnosis is later in childhood there may already be some effects of the disorder. Family, friends and carers will want to know how they can help. (Sample letters at the end of this chapter look at giving specific information to carers, preschool and school teachers, and others.)

As your child develops it will be important to talk about the diet and medications and the disorder with your child (see chapter 11).

Having 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 the metabolic disorder. (Chapter 1 explains how the disorder is inherited.) This is an individual decision, however the management of some of these conditions is so successful with early diagnosis that many parents feel there is no reason not to have another child.

Treating the metabolic disorder: the role of the family

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

Although it may seem overwhelming at first, your Metabolic Team will help you at each stage and in any way they can.

Successful treatment of these metabolic disorders includes the following:

1. A diet low in protein

The amount of protein allowed in your child's diet is calculated regularly. This is worked out by the Metabolic doctor and dietitian looking at your child's blood test results, age and growth.

2. A daily special formula or supplement

This is also calculated specifically for your child's needs. The formula or supplement should be taken several times throughout the day to maximize its effect. Additional vitamin and mineral supplements may be needed to make sure the diet is healthy

3. Vitamins or medications prescribed by your Metabolic doctor

The Metabolic doctor will advise you if these are needed to help keep the metabolic condition under control. You will be told how much to give and when, and the best way to give these at various ages.

4. Regular blood tests

Blood tests are the most accurate way to monitor how effective the diet and medications are. Your Metabolic Team will let you know how frequently blood samples are needed. This will vary from weekly to every three months.

5. Attending 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 monthly. As your understanding and confidence grows, the time between visits will increase. Most school-age children visit the clinic every three to six months. You will be able to attend more frequently if there are particular problems. Attending a Metabolic Clinic is also a requirement for receiving financial assistance from the government such as the low protein food grant (in Australia). See Chapter 16

'We ran out of formula one Christmas Eve and it was a nightmare to get supplies. Never again. When there are holidays such as Christmas or Easter just ahead I always place an order with my chemist two weeks before.'

6. Attending education sessions

Individual clinics may provide education sessions on managing a low protein diet (there are several other conditions that are managed in a similar way). 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. These sessions are sometimes held in conjunction with the support organizations, which can also provide support and education.

7. Keeping supplies up-to-date

Part of your responsibility is to make sure you have an up-to-date supply of prescriptions and blood test forms. It is important to keep at least a two week supply at home of supplements and medications as it can take a few days for the chemist to obtain them.

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

Metabolic Consultant

Metabolic Doctor on Call

Metabolic Dietitians

Metabolic Nurse

Clinic Booking Numbers

Sample letters for child care, preschool and school teachers and for parents of your child’s friends and classmates

The sample letters on the following pages can be photocopied and given to teachers, parents of children in your child’s class, and carers who need to know about your child’s metabolic condition. You can also use them as a basis for composing your own letters. Your Metabolic doctor will write to your doctor (G.P. and paediatrician) and give you copies of letters that you can show to health professionals involved in your child’s care.

When you are travelling overseas, taking a letter from your doctor explaining what the medicine is, how much is needed and when, and stating that is for personal use will save time and confusion with customs officials. It will also help if you leave the medication in its original packaging clearly labelled with the name of the person taking it and the instructions. See Chapter 15

Dear Teacher

………………………..has an inherited metabolic condition, called …………………………………………............ This means that his/her body is unable to break down protein in the usual way. This condition is a rare, non-contagious condition, which, left untreated, can result in damage to his/her health. Fortunately, the condition can be treated by a special diet and medications.

……………………… has been on this diet from birth (or since …. years of age). Apart from this 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 the usual school/preschool activities.

A metabolic condition is different from an allergy. If a child with this condition consumes food he or she shouldn't eat, there is no immediate reaction, but there will be a build-up of toxic substances and debilitating effects over time. The treatment for this metabolic disorder is a low protein diet, medicines and special nutritional supplements. We calculate the amount of protein from food 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

Please let us know in advance if you are having any special food events so we can arrange suitable alternatives. We will provide little cakes or lollies for substitutes for birthday cakes.

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.

If medications need to be taken to preschool/school we would like to discuss putting a system in place to make sure these are taken. If the special supplement is taken to school this also needs to be treated as a medication

Many thanks for your assistance. It is greatly appreciated. Please contact us on ……………………………………….if you need more information, or you can contact the Metabolic Clinic on …………………………………

Kind regards

Dear Parents

………………………. is a classmate of your child. She/he has a rare metabolic condition called …………………………………………..., which means her/his body cannot break down the protein from food in the usual way. This is an inherited condition she/he has had from birth, which was diagnosed at ………... of age. It is neither infectious nor contagious.

Treatment includes a low protein diet.

……………………………… 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 this metabolic disorder can't eat meat, fish, chicken, eggs, milk, soya products, beans or nuts. They eat a lot of fruit and vegetables, special prescription bread and pasta, and they take a special 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 a metabolic disorder eats the wrong food the problems happen over a longer 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 to a party, we'll be happy to talk to you about what food to give her/him. If your child is able to come to play at our house or to our child's party we will make sure there is food he/she would normally eat.

Kind regards

Phone:

Chapter 3

Homocystinuria, tyrosinaemia & hyperornithinaemia: Managing illness, pregnancy & health issues

Children with *certain different* metabolic conditions may get very sick with the usual childhood illnesses or accidents. They need extra special care at these times. This is why care during illness is mentioned throughout this handbook.

Homocystinuria, tyrosinaemia type 1 and 2, and hyperornithinaemia are NOT conditions that cause these extra problems with childhood illness. Illness may cause a temporary rise in the blood levels of the harmful metabolites in homocystinuria, tyrosinaemia and hyperornithinaemia, but this does not cause long term harm.

When you are unwell the body starts to break down its own tissues, releasing amino acids into the bloodstream. To prevent this, a high energy, low protein diet is generally recommended during illness.

When your baby is sick

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

Contact your local doctor or G.P. if your baby is unwell just as you would if they didn't have the metabolic disorder.

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 local doctor or G.P. as it is important to avoid dehydration.

When your young child is sick

Preschool children are likely to get between five and ten infections each year. Children and adults with these metabolic disorders get the same number of coughs, colds and other illnesses as other children.

If your young child is unwell:

- Give any medications recommended by your doctor.
- Encourage adequate fluid intake by offering drinks every hour or two while your child is awake. Fewer wet nappies or strong coloured urine is a sign your child may not be drinking enough.
- Dilute fluids and supplements – 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 – it is not necessary to have the full number of grams of protein intake. Return to the usual protein intake as soon as your child's appetite returns.
- Your dietitan may recommend you increase your child's energy intake and will explain how to do this.

Gastroenteritis

Listed below are general guidelines for children. These are also suitable for adults but not for babies. You should contact your local doctor if your baby has gastroenteritis or if your young child is not drinking.

- For the first 12 to 24 hours offer only oral rehydration solution

(ORS) or appropriate clear fluids – offer 100 to 200 ml every hour or two when your child are awake. See box next page.

- 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 protein intake. Appetite and a normal eating pattern will return as your child gets better.

Coping with sick days—older children and adults

For older children and adults follow these general guidelines for sick days (not gastroenteritis):

- Try to keep up an adequate fluid intake by taking a drink of cordial or juice every hour or two when 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 sick person's appetite without forcing and without aiming to include all the grams of protein allowance. Return to usual protein intake as appetite returns.

Medications

Most medications are suitable. Remember to tell a local doctor about the metabolic condition.

It is preferable with tyrosinaemia (but not necessary in homocystinuria or hyperornithinaemia) to avoid those medications sweetened with aspartame. If a medication is required urgently and contains this sweetener, give it until you can get an alternative or discuss it with your Metabolic doctor. Over the counter medicines should also be checked to see if they contain aspartame. There is more information on aspartame and tyrosinaemia at the end of this chapter.

Oral rehydration fluids

The following commercial products are available from chemists and need to be diluted according to the instructions on the packet:

- Gastrolyte ™(Aventis Pharm Pty Ltd).
- Pedialyte ™(Abbott)
- Hydralyte iceblock and ready made liquid ™(Splenda)

Note at the time of print some types of Gastrolyte contained aspartame. This is only of concern in tyrosinaemia (see above).

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

- dilute cordial (not diet): use 10ml + 150ml water
- Ribena™: use 10ml + 190ml water
- dilute soft drink or lemonade (not diet): use 50ml + 150ml water
- dilute fruit juices or fruit drinks: use 50ml + 150ml water
- sugar water: use 1 level 5 ml teaspoon sugar + 250ml water

Pregnancy

Homocystinuria, tyrosinaemia and hyperornithinaemia are rare so not many women with these disorders have had their own children. However a healthy baby can be expected as there does not seem to be any risk to the baby if you have these disorders and you keep well and healthy.

When you are **planning a pregnancy** talk to your Metabolic Team about the issues for you and your pregnancy and the risk that your baby may have the metabolic disorder (see Chapter 1 for how the disorder is inherited). There may be some changes you need to make or precautions that you should take.

It is important to:

- **Be healthy before you get pregnant**. Have your metabolic condition in good control and try to be in a healthy weight range. Check with your dietitian that your intake of vitamins and minerals is adequate before you get pregnant as well as during the pregnancy.

- **Maintain close contact with the Metabolic Team** before and during pregnancy. Let your clinic know as soon as you know you are pregnant. You will probably need more regular blood tests. Changes to your diet and amount of supplements will be necessary as your pregnancy progresses. You will need more protein in your diet as your baby grows and you may need to eat some foods you normally exclude to achieve this. The team will work with your obstetrician (pregnancy doctor) to provide the best possible care for you and your baby.
- Gain sufficient weight during the pregnancy (see chapter 14) but not too much. Talk to your dietitian about the best ways to achieve this and see chapter 14.
- Pregnancy in women with homocystinuria (but not the other disorders) increases the risk of blood clots, stroke and heart disease. Talk to your Metabolic doctor and obstetrician about managing your health during pregnancy and in the immediate time after the birth as the changes in your body during pregnancy and birth can increase your risk of complications. If you have homocystinuria you may be given blood thinning medication from the last few months of pregnancy until about six weeks after delivery.
- Breastfeeding should be possible with regular blood tests to check that your intake of protein and energy is adequate.

More information for those with homocystinuria

Eye testing

Short sight (myopia) and dislocation of the lenses is common in untreated homocystinuria. This is avoided or much delayed in early treated homocystinuria. Your Metabolic doctor will advise on how often eyes will need to be tested.

Bone density

Osteoporosis (thinning of the bones) and other bone problems are a problem of untreated homocystinuria. Good control of blood homocystine and methionine levels will reduce the risk.

Regular exercise and an adequate intake of calcium will also help. Your dietitian can assess your calcium intake because low protein diets can be low in calcium.

Reducing the risk of thrombosis (blood clots)

There is a slightly increased risk of thombosis (blood clots) when you have homocystinuria. Your doctor will advise you about what to do at times when the risk of these is high, for example when having surgery, when traveling by aeroplane, during and after pregnancy.

General advice to reduce the risk:

- Keep blood levels of homocysteine and methionine under good control and take all recommended supplements and medications.
- Maintain a weight that is healthy for height – your clinic will advise. Also see Chapter 10.
- Eat healthy fats and reduce your intake of salt for heart health – see Chapter 10.
- Exercise regularly.

Air travel

When travelling by air the risk of clots in the large veins is increased. The following will reduce this risk:

- Drink plenty of (non-alcoholic) fluids during flights.
- Move the ankles and massage the calves. Do the exercises recommended on in-flight entertainment or in the magazine.
- Wear loose, non-restrictive clothing and flight socks or stockings.
- Take care moving around the cabin—the risk of injury from turbulence may outweigh the benefit of the exercise.
- Be vigilant for the symptoms of a deep vein thrombosis, in particular pain in the calves, during and for up to a month after long flights. If symptoms occur, seek medical advice without delay.

(This information is from the Australasian Society of Thrombosis and Haemostasis 2003 and recent research on benefits of flight socks.) There is more general advice on overseas travel in Chapter 15

Surgery

Talk to your Metabolic doctor if surgery is planned. The team will advise your surgeon on your particular needs.

More information for those with tyrosinaemia

Some artificial sweeteners used in food and medications contain phenylalanine and should be avoided. To find out whether a product contains these artificial sweeteners check the *ingredient list* on the food label.

Additives 951 and 962 (also called Nutrasweet, Equal, Canderal and aspartame-acesulphame) contain aspartame which is a source of phenylalanine. 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
951 Aspartame	950 Acesulphame
962 Acesulphame-Aspartame	952 Cyclamate
962 Acesulphame-Aspartame	953 Isomalt
Nutrasweet, Equal, Canderal are other names used	
	954 Saccharin
	955 Sucralose
	956 Alitame
	957 Thaumatin
	961 Neotame
	965 Maltitol
	966 Lactitol
	967 Xylitol
	968 Erythritol
	Mannitol, sorbitol

Diet drinks

Nutrition Information

Servings per package: 3
Serving size: 200ml

	Quantity per serving	Quantity per 100g (or 100ml)
Energy	4 kJ 1 Cal	2 kJ 0.5 Cal
Protein	0.1g	0.05g
Fat, total	0 g	0 g
- saturated	0 g	0 g
Carbohydrate	0.15g	0.07g
- sugars	0 g	0 g
Sodium	20 mg	10 mg

These drinks contain very little protein but do contain phenylalanine so should be avoided by people on a diet for tyrosinaemia. 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 phenylalanine in the aspartame. So along with the nutrition table you will 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 aspartame or aspartame-acesulphame added, it is recommended that you advise your metabolic doctor.

When buying over-the-counter medications always check whether they contain aspartame or aspartame-acesulphame. You can ask the chemist about the sweeteners in the medication. If they are uncertain they should contact the supplier to check it for you.

Chapter 4

Blood samples

About blood samples

Blood samples are needed to check blood levels of amino acids or other blood components, such as ammonia, affected by the metabolic condition. The results of these blood level tests are used to change the diet and other treatments to best manage the condition at any one stage.

For some metabolic conditions, the blood levels of amino acids or other components can be measured from blood collected on a special type of card, similar to that used for the newborn screening program. The blood is taken from the heel or finger and the card mailed to the testing laboratory. If this is possible you will be shown how to do this yourself. Pages 5 and 6 give more information on collecting blood in this way

Other blood level tests may need to be done by a pathology service or laboratory. You may be able to do this close to your home and the samples will then be sent to the testing laboratory.

How often blood samples are 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 daily, 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, for some metabolic conditions weekly, fortnightly or monthly tests continue throughout life.
- Additional blood samples may be needed after and during an illness or when the levels are too high or too low.

Blood testing is the only accurate way to monitor control of metabolic conditions. Tests are taken regularly—your Metabolic Team will let you know how often.

Getting the results

Blood test results are sent to the Metabolic Team and/or the regional dietitian, paediatrician or local doctor (G.P.), 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 metabolic conditions.

What affects blood test results

Different blood tests are used for different metabolic conditions. Some information is given in Chapter 1 and your Metabolic doctor can explain what is useful in the particular condition.

Some general points about blood test results:

- It is normal for most blood results to have a range of acceptable results, rather than one ideal number.
- Your clinic may advise you to have blood taken at specific times of the day e.g. not just after a feed or meal. Results can be different at

different times of the day.
- Amino acid levels in the blood can go too high or be too low. Too high means the body has more than it needs at that time. Too low means more is needed for building protein within the body.

High levels of amino acids or harmful metabolites may occur because:
- Intake of protein from food is too high.
- Rate of growth has slowed so that for now protein intake is too high.
- Sickness.
- Not eating enough food or taking enough of the special supplement so that the body breaks down its own tissues for energy. This can happen if there is a major upset in your child's life as well as during sickness.
- Not taking medications prescribed by the metabolic doctor or a need for a change in dose.
- Weight loss – intentional or not. If needed, it is best to try and lose weight very gradually to maintain good blood levels.
- Levels of training for sports have been reduced e.g. training for rugby, athletics or swimming squad is over. (High levels of activity increase muscle growth and increase protein needs).

Low levels of amino acids may occur because:
- Intake of protein is not high enough.
- There is an imbalance between the protein from food and the amount of the special supplements.
- A growth spurt in children or weight gain is using up amino acids to build new tissue.
- Additional activity such as regular gym workouts or training for seasonal sports.

If blood levels are too high or too low, the Metabolic Team will discuss ways to bring them under control.

Involving children in their blood tests

Blood tests are a fact of life for children with metabolic conditions. 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.

Even relatively young children can have some role in collecting their blood samples. You can involve your child by:

- Letting toddlers decide which arm or finger to use for the blood sample.
- Making sure they are warm and encouraging them to do circles with their arm to get the blood flowing.
- Giving 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 into the tube or onto the card.
- Telling them the blood will be taken on the count of three, and get them to count with you so they know when to expect it.
- Giving lots of positive feedback about how well they have done.
- Avoiding reinforcing any negative messages—phrases such as "isn't the lady cruel for making your arm hurt" or "isn't daddy naughty for making your finger sore" links the blood test with punishment.
- As soon as the child is old enough, encouraging them to introduce themselves to the pathology service, write their name on the card or push the button on the diabetic pen. Involving the child in the process helps make it an achievement rather than an imposition.
- Making use of favourite theme sticking plasters. Get young children to help press on the cotton wool over the puncture site.
- Being 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 – fear of the unknown is very real in children.

How to take a card sample from the heel or finger

Parents are taught how to collect samples from babies and young children. This usually occurs within the first few weeks. Nursing staff or other health professionals will take the samples until you feel you're ready to learn how to do it yourself. In New Zealand, during the first four to 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 blood collection cards (also called Guthrie cards) 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).

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. At the laboratory, small circles of blood are punched out of the card and tested. A single layer of blood is required for the test to be carried out accurately. For this reason it is important that the blood completely soaks through the card and that blood drops are not placed on top of each other.

1. Fill in the details on the special blood collection 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')
- blood collection card (Guthrie card)
- tissues or cotton ball
- sticking plaster

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 an area to take the blood sample from either his hand or heel. Heel samples are best taken towards the side of the bottom of the foot not in the centre. Ask your metabolic nurse to show you.

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 sticking plaster 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 that 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*, depending on which type of card you have, either use the wrap flap on the card or wrap the card in plain paper. The Metabolic nurse or another health professional will demonstrate this to you.

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).

Taking a finger prick sample

Until a child is at least six months of age, blood samples are taken from the heel. You can then change to fingertip samples or wait until the children starts walking. 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, such as a diabetic ‘pen’, available from pharmacies. The best ones have a dial that controls how far the skin is penetrated. You can 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. Or you may wish to start at the highest setting and work downwards.

The sample can be taken from any finger, but is best taken from the middle and fourth fingers. Generally the side of the fingertip bleeds better than the pad and is less painful.

Trouble shooting card samples

Common problems with samples, which generally mean the laboratory is unable to determine an accurate blood level, include the following:

- blood spot is too small
- one drop of blood has been put on top of another
- blood has not soaked through to the back of the card
- card has got wet
- foot or finger has been squeezed too hard – no squeezing or gentle squeezing is best
- 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 with card samples

‘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 the blood test is associated with fun.’

The following may also help:

- Make sure the hand or foot is warm before taking the sample—you can do this 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 he or she 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.

Where card samples are sent

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

Australia
Blood collection cards in Australia are sent to the newborn screening laboratory service attached to your Metabolic clinic. The address is on the card. There are five screening laboratories:

- New South Wales which also covers the Australian Capital Territory
- Victoria which also covers Tasmania
- Queensland which also covers part of the Northern Territory.
- South Australia which also covers part of Northern Territory
- Western Australia

Chapter 5

Understanding the special formula or supplement

THE FIVE PARTS OF THE LOW PROTEIN DIET

- Special formula or supplement (see below)
- Counted protein from food (see Chapter 6)
- Low protein foods (see Chapter 7)
- Meal planning and healthy food choices (see Chapter 10)
- Changes needed during illness (see Chapter 3)

People who are not on a low protein diet get all the protein they need from the food they eat. When protein is restricted to treat a metabolic condition, the special formula or supplement makes up for nutrients they don't get from food. **The special supplement is vital. Children with some metabolic disorders can get very sick if they don't take it.**

Different kinds of supplements

The special supplement (which, for babies, is called a special formula) plays an essential role in the low protein diet. Your Metabolic Team will prescribe the most appropriate formula or supplements.

There are two types of formula or supplement which may be used depending on the metabolic disorder and the amount of protein restriction needed:

- **Protein supplements**. These provide the other amino acids which are not affected by the metabolic disorder. This helps the body make protein and helps keep the metabolic condition in good control. These also provide energy (calories or kilojoules) as well as vitamins, minerals and trace elements essential for a healthy, balanced diet.

- **Protein free supplements**. These provide energy (calories or kilojoules) from carbohydrate and fat. It is essential for good control of the metabolic condition and for growth that your child takes in enough energy on a low protein diet. These supplements usually also contain vitamins, minerals and trace elements essential for a healthy, balanced diet.

'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'

Your doctor or dietitian may prescribe vitamin, mineral or individual amino acid supplements as well if your child's (or your) diet does not contain enough vitamin and minerals or blood tests show levels of individual amino acids are low. This is to make sure the diet is healthy and balanced. This is different from the large doses of some vitamins that may be prescribed by your doctor to treat some metabolic conditions.

Protein supplements

Protein supplements are made to suit the nutritional needs of the various metabolic conditions and people of different ages. They 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 a special formula; toddlers, older children and adults have more protein supplement options which come in the following forms:

- **Powder** which 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 may need to be taken as well.
- **Gel or thick paste** which can be eaten with a spoon or taken as a low volume drink – available flavoured or unflavoured.
- **Liquids** – ready made liquids are available for some metabolic disorders.

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

'I know I have to take my supplement but having a choice does make it easier'

The Metabolic doctor (or possibly your local doctor/ GP) will provide a prescription for the supplement, which is available from pharmacies. As metabolic disorders are rare conditions 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 makes the product.

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

Protein free supplements

A protein free formula (Energivit™, Pro-Phree™) is used in some metabolic conditions to provide energy (from carbohydrate and fat), vitamins, minerals and trace elements essential for a healthy, balanced diet. Increased amounts may be needed during illness. Other supplements may be used by older children to provide additional energy (from carbohydrate and/or fat) e.g. Duocal™, Polyjoule™ or Polycose™, Polycal™. Some of these supplements are available on prescription; some need to be purchased.

Vitamin, mineral or individual amino acid supplements

Your clinic doctor or dietitian will advise you if these are needed and how to obtain them.

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 some supplements. Alternatively you may like to use a home delivery service if this is available.

Always check the product supplied exactly matches the one prescribed by your Metabolic Team.

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

Taking the special supplement

It is best to take the supplement with meals in several doses spread throughout the day – ideally over at least 12 hours. This is easy with babies who feed frequently but can be more of a challenge for older children and adults.

'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.'

Also:

- follow the supplement prescription carefully
- if you weigh your supplement, check that your scales are accurate – do this by measuring 50ml of water and weighing it, it should weigh 50g
- if you use tablespoons to measure your supplement, make sure you are 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 not rounded scoops or tablespoons.

Tips for parents of young children

The full amount of the special supplement prescribed should be taken every day over several doses. This is important for growth and development, and maintaining the right blood levels of amino acids or ammonia or other harmful metabolites. Follow these tips on including the supplement:

- Treat it 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.
- Give the supplement 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.
- Divide the daily volume into three or four small amounts that will not overwhelm your child.

- Remember that the supplement is acidic and could result in damaged teeth if your child sips on it often throughout the day (see Chapter 10 for advice on dental care).
- Give it to your child in a sipper cup or cup rather than an infant formula bottle starting from when your child is about twelve months old.

Right from the beginning, talk to your child about their metabolic disorder. 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 your child know **taking it is not negotiable.** Make sure all the supplement is drunk, including the sediment at the bottom, which contains important minerals and vitamins.

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 essence, peppermint essence, or tomato juice.

When your child refuses the supplement

If your child is having difficulty drinking the prescribed dose of supplement on a regular basis, discuss this with your dietitian. There may well be some things you can do to make it less of a battle. Some reasons for your child refusing the supplement can be that:

‘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’.

- your child is snacking a lot or having other drinks - instead of drinking the supplement
- 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.

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

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

'When we go camping I carry bottled water that I can keep cold. Before we go, I weigh half-day portions into zip lock bags. This works for us. We pull out a bag of powder, add water and pour into a cup, rinse out the shaker and it's ready for washing up.'

Taking the supplement when away from home

If you have young children take the supplement with you for your child to drink with the meal, when you are eating out, If this is not practical, give some before and after the meal. Use insulated carry bags or small thermos containers to keep drinks cool.

At school and work

Encourage your child to take some 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 dinner time.

' I always take extra supplement with me when I'm away from home in case of an unexpected delay, such as car trouble – even if I'm only going half an hour down the road.'

When you take your supplement to work, if you prefer it chilled and there's no fridge available, 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 chapter 16 and in the quiz (Chapter 19). 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 at the end of Chapter 15. It is also recommended that you keep the supplement in its original containers with the labels in tact.

Chapter 6

Counting protein from food

The low protein diet

THE FIVE PARTS OF THE LOW PROTEIN DIET

- Special formula or supplement (see Chapter 5)
- Counted protein from food (see below)
- Low protein foods (see Chapter 7)
- Meal planning and healthy food choices (see Chapter 10)
- Changes needed during illness (see Chapter 3)

People on a low protein diet take in less protein than the average Australian or New Zealander does, but it is enough for growth and functioning of the body. Sometimes additional protein may be provided by the special supplement.

The low protein diet consists of:

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

Foods such as meat, chicken, fish, eggs, milk, yoghurt, cheese, soya products, nuts and legumes (e.g. lentils, chick peas, kidney beans) are usually too high in protein to include. Some people may be able to include small amounts of these foods. You should discuss this with your dietitian.

See the diagram, **Five Food Groups** in chapter 1 which compares a protein restricted diet with usual intake.

The amount of protein needed and tolerated by each person, even with the same metabolic disorder, is quite different. The diet is adjusted according to individual blood test results and growth, and is likely to vary from time to time.

How the protein intake is counted

Foods such as cereal based foods and some vegetables and fruits contain small amounts of protein. These foods have to be counted in the diet. To do this you will need protein food exchange lists, which your dietitian will provide. This shows how much protein is contained in common foods, 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’ – that is foods that contain little or no protein which do not need to be counted.

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 the information you need to decide whether that food can be included. If the diet is for tyrosinaemia you will also need to check the ingredient list for certain sweeteners (see chapter 3).

Reading the nutrition information table

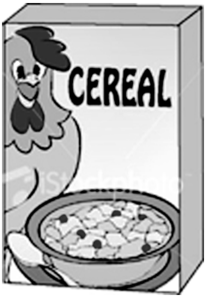
One of the things you need to check when choosing food for a low protein diet is the nutrition information table. This tells you the amount of energy (kilojoules or calories), protein, fat, carbohydrate, fibre and other nutrients the food contains.

The table also tells you how much protein the product contains per serve and per 100g. You can use this to decide whether to include this food in the low protein diet. 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 your child’s condition and the way you count protein. Be guided by your dietitian about what you can use ‘freely’ in the diet.

How to calculate protein from food labels

Breakfast rice pops



Nutrition Information		
Servings per package:20 *Serving size: 300g (¾ metric cup*		
	Quantity per serving	*Quantity per 100g*
Energy	460 kJ 110 Cal	1530 kJ 367 Cal
Protein	1.4g	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

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

Using a calculator and following the information on the back of the packet:

100g Rice Pops = 4.6g protein
20g 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 this straightforward. When in doubt talk to your dietitian. Here are some examples of labels that can cause confusion:

- **Protein content per serve is <1g (less than 1g)**: when foods contain less than 1g 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 26 Cal	1385 kJ 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 = 192g

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

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

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

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

- **Products that give values for the product mixed with water**: If you don't read the nutrition information table carefully it is easy to be fooled into thinking the food is much lower in protein that 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 100g prepared cereal*
Energy	80 kJ 18 Cal	170 kJ 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 45g serving of baby rice is actually 5g of the dry rice cereal and 40ml water. All of 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, you need to remember that 5g dry cereal contains 0.4g protein.

Chapter 7

Low protein foods, recipes, cooking & shopping

THE FIVE PARTS OF THE LOW PROTEIN DIET

- Special formula or supplement
- Counted protein from food (see Chapter 6)
- Low protein foods (see below)
- Meal planning and healthy food choices (see Chapter 10)
- Changes needed during illness (see Chapter 3)

Low protein foods are an important part of a low protein diet. They add energy and variety and allow foods and meals to be prepared that are similar to those the family is eating. Chapter 10 gives more information on planning meals and snacks for a low protein diet.

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 low protein recipes, and inventing new ones as they gain experience. Be adventurous, experiment, and share your successes with other families.

> Phenylketonuria (PKU) is one of the more common metabolic conditions affecting protein metabolism. Recipes for PKU are usually suitable for low protein diets but you will need to count protein in the way you have been taught.

Here are some basics to get you started cooking the low protein way. You will also find that support organizations or websites are a good source of new food ideas and recipes (see resources chapter 17). Ask your dietitian for more tips, recipe books 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 125g low protein flour for cakes.
- Add 1½ teaspoons baking powder to 125g 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 (check protein, brands vary and some have added chickpeas) cream (check protein), margarine mixed with water (100g margarine and 100ml water beaten together), coffee whitener, Vance’s Darifree™, Milupa lp drink ™(count the protein) or Loprofin PKU Milk Tetra Pack™. You can use these to make custards, desserts and white sauce.

Tips for low protein cooking

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 (beat) 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. Adjusting the volume slightly can make a big difference.

PASTRY & BISCUITS

- Chill pastry and biscuit dough 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.

Low protein bread recipes

Note; Metamucil™ used in these recipes contains fibre from psyllium husks and is available from chemists

WHITE BREAD

3¼ cups low protein flour
1 tablespoon sugar
1½ cups warm water + 3 tablespoons
1 tablespoon + 1 teaspoon Metamucil™
¾ teaspoon salt
1¾ teaspoons yeast (8g packet)
3 tablespoons coffee whitener

Method:

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

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 (½ 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 USING BREAD MAKER

400g low protein flour
2½ tablespoons Metamucil ™ (regular variety)
1½ teaspoons active dry yeast (8g)
1 tablespoon sugar
1 teaspoon salt
1 tablespoon vegetable oil
380–400ml warm water

Method:

- Sift flour twice into a bowl.
- Add Metamucil ™, sugar, salt and yeast and mix thoroughly.
- Add dry mixture to bread machine pan.
- Add the oil and water and start the machine straight away.
- 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 USING SMALLER BREAD MAKER

300g low protein flour
1 teaspoon active dry yeast (5.5g)
4 teaspoons Metamucil ™
2 tablespoons coffee whitener
1 teaspoon salt
280ml water

Method:

- Sift flour twice into a bowl.
- Add Metamucil ™, yeast, coffee whitener and salt and mix thoroughly.
- Add dry mixture to bread machine pan.
- Add water and start the machine straight away.
- 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

2 cups (290g) low protein flour
250ml water
⅛–¼ teaspoon yellow food colouring
favourite herbs or spices to taste

Method:

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

RUSKS

1 cup low protein flour (145g)
⅓ cup water

Method:

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

PIZZA

Serves 4

Base
225g low protein flour
pinch salt
2 teaspoons baking powder
50g margarine
150ml water (approximately)
Topping
Basic topping
25g margarine
1 onion, chopped
pinch mixed herbs
black pepper
OR use commercial tomato based pizza topping or pasta sauce

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

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

Potato topping
70g sliced baked potato
60g sliced baked sweet potato

60–90g grated low protein cheese per pizza (optional)

Method:

- Sift together the flour, salt and baking powder.
- Rub in margarine with fingertips then add enough water to make soft dough.
- 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.
- Spread the basic topping plus one of the other toppings on each pizza. Sprinkle with cheese (optional).
- Bake at 180°C for 30–35 minutes until the base is cooked and the topping is browned.

POTATO BALLS

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

Method:

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

Low protein cakes, biscuits and scones

DATE SCONES

1 ⅔ cups low protein flour (200g)
pinch salt
1 teaspoon baking powder
30g margarine
60ml cream
20g castor sugar stirred into 100ml warm water
30g chopped dates

Method:

- Preheat oven to 200°C and grease an oven tray.
- Sift flour, salt and baking powder together into a bowl.
- Rub margarine in with fingertips.
- Add cream and water, then chopped dates and mix to a soft dough.
- Turn onto a floured board and knead lightly.
- 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.
- Cook for 20 minutes on the middle shelf of the oven.
- Store in the freezer or in an airtight container in the fridge.

SPONGE CAKE

This is a very easy sponge to make. You can add extras (such as grated rind and juice half lemon) for variety. This basic recipe can be used to make special occasion cakes, and baked or steamed sponge puddings.

200g low protein flour
110g margarine
110g castor sugar
2 teaspoons baking powder
2 teaspoons egg replacer
150ml water
1 teaspoon vanilla essence (or to taste)

Method:

- Preheat oven to 180°C.
- Place all ingredients in a mixing bowl and beat well for two minutes using an electric mixer, processor or wooden spoon, until the mixture is light and fluffy and has a soft, dropping consistency.
- Place in one 20 cm (8 inch) tin or divide into two greased 18 cm (7 inch) tins.
- 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.
- Remove from the tin(s) and cool on a rack.
- Sandwich together with jam or other filling. Dust with icing sugar. Store in an airtight container.

CUP CAKES

Place 2 teaspoons of the sponge cake mixture into each paper patty case and cook for 6 to 8 minutes.

SWEET BISCUITS

From Special Recipes *by Eileen Green, The West Midlands Support Group for Phenylketonuria (out of print).*

100g margarine
100g castor sugar
1 teaspoon egg replacer
few drops vanilla essence
200g low protein flour
1½ tablespoons water

Method:

- Preheat oven to 180°C.
- Beat together margarine and sugar until light and fluffy.
- Mix in vanilla and egg replacer.
- Gradually add the low protein flour and stir well. Add water and mix to form dough. Use your hands if you find it easier.
- 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.
- Cut into shapes using biscuit cutters.
- Place on a lightly greased oven tray and cook 15-20 minutes or until the biscuits are just turning brown.
- Cool on a wire rack. These keep well in an airtight container.

Try these 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 50g 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.

Low protein muffins, pancakes, crumpets and pikelets

APPLE MUFFINS

Makes 6 muffins

1 cup low protein flour
3 tablespoons sugar
2 teaspoons baking powder
½ teaspoon baking soda
½ teaspoon cinnamon
¼ teaspoon salt
3 tablespoons vegetable oil
⅓ cup water
⅓ cup (80g) apple puree

Method:

- Preheat oven to 200ºC.
- In a medium mixing bowl, stir together flour, sugar, baking powder, baking soda, cinnamon and salt.
- 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).
- Spoon batter into six greased 6cm muffin pans (batter tends to stick to cupcake liners). Bake for 15–18 minutes.

PANCAKES

Makes 4 pancakes

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

Method:

- Mix the flour and egg replacer with 3 tablespoons of liquid to form a paste.
- Gradually work in the remaining liquid to make a smooth batter with pouring consistency.
- 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.
- Allow to cook for a few minutes and when the underside is done, flip the pancake over to cook the other side.
- Repeat with the rest of the batter.

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

Makes about 12

2 cups low protein flour
1 tablespoon baking powder
2 tablespoons custard powder
1 teaspoon sugar
½ teaspoon salt
2 cups warm water

Method:

- Mix the dry ingredients.
- Add water and mix to a smooth batter (about two minutes with electric beaters).
- Allow to rest for 10-15 minutes.
- Lightly grease frying pan and heat.
- Grease egg rings.
- 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).
- Do not brown them too much to allow for toasting.
- Leave for seven to ten minutes to cool and dry.

QUICK SAVOURY PIKELETS

Makes 8

½ cup low protein flour
30g chopped onions
¼ teaspoon baking soda
1 teaspoon chopped parsley
100ml water
¼ teaspoon mixed herbs
pinch pepper
oil for frying

Method:

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

Low protein custard recipes

VANILLA CUSTARD

Serves 2

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

Method:

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

Try these variations:

Blancmange Use ¾ cup water only. Follow the same method as for the custard and pour into a wet decorative mould.

Velvet Replace 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

⅔ cup water (165ml)
⅓ cup rich cream (85ml)
1 tablespoon custard powder (10g)
2 teaspoons sugar (10g)

Method:

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

Finding recipes elsewhere

Cook books catering for people on a low protein diet are few and far between, but many recipes can be easily adapted for the low protein diet. Most recipes for a phenylketonuria(PKU) diet are suitable but you will need to count the protein as you have been shown in chapter 6.

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 are another great source of ideas. These are regularly printed in *PKU NZ Newsletter*, *MDDA newsletter* and *NSW PKU Association Journal* and on the websites for MDDA, the PKU Tree and SHS:

http://www.mdda-australia.org/
http://www.pkutree.com.au/
http://www.shsweb.co.uk/metabolic/pat/
http://www.shsweb.co.uk/metabolic/pkubook/

Other low protein and PKU recipes can be found on the web at:
http://www.cambrookefoods.com/fsRecipes.htm
http://www.dietspec.com
http://www.ener-g.com/recipe/

(not all the low protein products used in these recipes are available in New Zealand and Australia but you can adapt the ideas).

Cook books with recipes for a low protein diet

- *Low Protein Cook Book Recipes for Life* by Jo Boston, 2007. This book has been written by a New Zealand mum who has a child with a protein disorder – ask your Metabolic dietitian for an order form.
- *The Low Protein Collection*, published by SHS Nutrition Services
- *Special Recipes* by Eileen Green, West Midlands Support Group for Phenylketonuria (out of print – your clinic may have a copy)
- *Variety in Life for PKU— a recipe book*, published by SHS International Limited (out of print but your clinic may have a copy)

Cook books with easily adapted recipes

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

If weight loss is a priority, low fat cook books 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 cook books on their shelves. Children’s party cook books 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 the 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 protein for vegetables such as potato, sweet potato, corn and peas, then do the same when they’re in a recipe. Adapt the recipes according to how strict a low protein diet you are following.

How to adapt a recipe

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

- leave out the meat, chicken, fish, legumes and nuts

- use a low protein milk substitute for milk, e.g. rice milk, Duocal ™, coffee whitener, Vance's Darifree™ and Milupa lp drink ™
- where a recipe includes a white sauce, use your own favourite low protein sauce recipe or a packet mix that is low in protein
- use sour cream instead of yogurt
- choose a low protein stock
- use egg replacer instead of egg
- instead of cheese in risotto use a small amount of grated low protein cheese or stir in a spoonful of coffee whitener, Vance's Darifree™ or Milupa lp drink ™ powder
- use milk-free margarine or oil instead of butter or margarine containing milk if that is what your diet requires
- use cornflour or low protein flour instead of plain flour if that is what your diet requires
- use low protein pasta or low protein rice instead of regular pasta and rice if that is what your diet requires.

General cook books

The following cook books are sold through newsagents and bookshops and will contain some recipes you can use or adapt. In the future they may be replaced by updated versions with different titles. If you cannot find these books look for similar titles, or check at your local library. Some of these may be out of print.

This is a new list put together by one of the dietitians:

- *Australian Women's Weekly New Salads,* Ed. Susan Tomnay et al, Sydney. ACP Books , Sydney, 2006
- *Salads Simple Fast and Fresh***,** *Australian Women's Weekly Home Library,* Ed. Susan Tomnay et al, Sydney, ACP Publishing, Sydney, 1999
- *The essential vegetarian cookbook,* Ed Wendy Stephen et al, Millers Point, NSW, Murdoch Books, 2004
- *Australian Women's Weekly Vegie Food***,** Pamela Clark, Sydney, ACP Publishing, 2006
- *Australian Women's Weekly Fast Vegies,* Pamela Clark, ACP Publishing , Sydney 2007
- *Australian Women's Weekly Fast Pasta***,** Pamela Clark, Sydney, ACP Publishing, Sydney. 2007

- *The essential pasta cookbook*, Ed Wendy Stephen, Millers Point, NSW Murdoch Books, 2004
- *The essential rice cookbook*, Ed Zoe Harpham, Millers Point, NSW, Murdoch Books, 2004.
- *Australian Women's Weekly Soup*, Pamela Clark, Sydney, ACP Publishing, 2007
- *Australian Women's Weekly Great Italian Food: traditional and modern dishes***,** Sydney, ACP Books, 2006.

Shopping list

The low protein shopping list includes regular supermarket items and special low protein products.

Always check the nutrition labels on the packets to find out the protein content of each serving. How much can be eaten will depend on the protein allowance for the day.

The special low protein products are identified with an asterisk (*). In Australia special low protein products can be ordered directly from the supply company (order forms are available from your dietitian). In New Zealand, many are available on prescription, which your doctor will provide, others can be ordered directly from the company. The Loprofin, Milupa and low protein cheese products listed below can be ordered from Nutricia; the Aproten and Agluten products from Sharpe Laboratories

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

Breakfast cereals

☐ Loprofin Breakfast Cereal Loops™*
☐ Kellogg's Coco Pops™
☐ Kellogg's Fruit Loops™
☐ Kellogg's Honey Crispix™
☐ Kellogg's Rice Bubbles™
☐ Kellogg's Corn Flakes™
☐ Sanitarium Ricies™
☐ Sanitarium Weetbix™

Bread

- ☐ Loprofin Sliced Loaf™*
- ☐ 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, lasagne*
- ☐ Loprofin™ macaroni, pasta spirals, spaghetti, lasagna*
- ☐ 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 starch flour
- ☐ cellophane noodles
- ☐ fresh rice noodles
- ☐ Loprofin™ rice
- ☐ Agluten riso™ *
- ☐ rice – any variety
- ☐ couscous, polenta, semolina
- ☐ sago (seed tapioca), pearl tapioca

Biscuits and crackers

☐ Aproten™ low protein cracker toast, crispbreads*
☐ Loprofin™ Herb Crackers, Savoury Crackers*
☐ Loprofin™ sweet biscuits* (Nutricia)
☐ Orgran™ Rice crispbreads, Orgran corn crispbreads
☐ Arnott's Cruskits™, Rice Cruskits™,
☐ cracker bread
☐ rice cakes
☐ rice crackers
☐ corn cakes, corn thins
☐ Arnott's SAOs™,
☐ Paradise Lites
☐ Arnott's™ Jatz, rice jatz
☐ Arnott's™ Saladas, Premium

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

Snack foods

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

Ready prepared sauces

☐ tomato paste, tomato puree, tomato-based pasta sauces
☐ ready made sauces or recipe bases including pasta sauces, white or cheese sauce mixes and casserole bases – check the nutrition labels on the many varieties available.

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.

Dried packet soups – choose vegetable based soups, such as tomato.

Alternatives to dairy products

☐ coffee whitener
☐ rice milk
☐ Vance's Darifree ™
☐ Milupa lp drink ™*
☐ Loprofin PKU Milk Tetra Pack™*
☐ low protein cheddar cheese*
☐ low protein mozzarella cheese*

Miscellaneous

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

Chapter 8

Feeding your baby

Breast and bottle feeding

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.

> Infants with disorders affecting protein metabolism can be successfully breastfed and keep blood levels of the harmful metabolites at a safe level. Breast milk contains much less protein than infant formula. However, breast milk alone contains too much protein for some babies with metabolic disorders, who need a low protein diet. For these babies a special formula is given by bottle to take the edge off your baby's appetite before breastfeeding.

If you choose not to, or are unable to breastfeed, standard infant formula is the appropriate substitute, along with the special formula. It can be difficult to maintain a supply of breast milk particularly if your baby has been unwell prior to diagnosis.

The special formulas contain all the nutrients needed for growth, except protein or the specific amino acids your baby has difficulty metabolising. Your Metabolic Clinic will prescribe the appropriate formula for your baby.

Your baby's special formula is called……………………………….

Your newborn's first days

For the first few days your baby may be given just the special formula) and no breast milk or standard infant formula. This will allow high blood levels of harmful metabolites to come down more quickly.

The clinic will advise you how much special formula your baby is likely to drink and how to make it up. (For guidelines see **Preparing, storing and feeding formula** later in this chapter.) Babies usually make the change to the special formula quite easily.

If you are breastfeeding, you will need to express to keep up your supply of breast milk. (See the section **How to express breast milk** later in this chapter.)

Re-starting breastfeeds and standard formula feeds

After the first few days expressed breast milk, breastfeeds or standard formula are introduced. Your baby will take less breast milk or formula than before because he is drinking a measured amount of special formula. This will reduce baby's protein intake. Most babies quickly learn to cope with the combination of bottle and breastfeeding or the two different formulas.

The Metabolic Team will work out a feeding plan for your baby. The order in which you give the special formula and breastfeed or standard infant formula is important, but may change from time to time. Usually the breast is offered after the special formula has been given. Formula-fed babies are usually given the special formula last or the special formula and standard formula may be mixed together. The Metabolic doctor or dietitian will tell you how much to give and what to do if your baby seems hungry

The amount of special formula and breast milk or standard infant formula will need to be adjusted from time to time to provide the right balance to meet your baby's needs and keep blood test results in the 'safe' range.

Regular measurements of weight, length and head circumference are taken to make sure your baby is growing well. This can be done at the clinic or at your local health centre and the results will be reviewed by the Metabolic Team. Appropriate gains in weight and length and plenty of wet nappies are re-assuring as these show your baby is getting enough milk and formula.

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

Breast milk supply

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 test results are in the safe range, your baby is getting the amount they need. After the bottle of special formula, alternate the breast you offer first. This will help maintain your supply. If your baby is not interested in the second breast, don't worry, offer this one first at the next breastfeed. If you are concerned you can increase your supply by expressing breast milk three or four times a day after your baby has breastfed. See sections **How to express breast milk** and **Looking after Mum** later in this chapter.

Preparing, storing and feeding formula

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 **Cleaning and sterilising** later in this chapter)
- use cool boiled water to make up feeds (empty the kettle, refill it and allow it to boil, pour into sterilized bottles and allow to cool)
- measure the water and formula accurately

- unless your clinic team advises you to do so, do not add anything to your baby’s formula.

To store the formula:

- ideally make up each bottle as required
- if you make up enough for several feeds, place it in the coldest part of the refrigerator, usually at the back, as soon as it is made
- throw out any leftover feed after 24-hours.

At feed time

- Carefully measure the amount of formula required into a feeding bottle.
- 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. Many babies will happily take cold formula even straight from the refrigerator.
- 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.
- Hold, cuddle and talk to your baby while he is being fed.
- If your baby doesn’t drink all of the formula, record what has been drunk and throw out what is left after an hour.

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 powder 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 babies 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.

Many young babies enjoy extra short snack feeds to settle. If breastfeeding, your baby may have extra breastfeeds in addition to the special formula and usual breastfeeds, as long as all the prescribed amount of special formula is drunk in 24 hours. If baby is fully bottle fed you can give extra of the special formula.

A new feed is considered to start when an hour or more has passed since the last feed finished.

Provided your baby is well, nothing apart from special formula and breastfeeds or special formula and infant formula are 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 unless prescribed by your doctor.

How to express breast milk

If your baby is not able to breastfeed, which may happen for a few days after diagnosis or if breastfeeding is interrupted at any other time, 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 if you are not breastfeeding at all. If you want to increase your supply of breast milk you can express three to four times a day after baby has breastfed.

- First, wash your hands.
- 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.
- If you're using a pump make sure the pump parts that come into contact with the milk are sterile.
- Your milk can be stored in bottles and frozen for later use. It will keep for two weeks in the freezer section inside a refrigerator, three months in the freezer section of a refrigerator (with a separate door), or six to twelve months in a deep freeze.
- 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.
- 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 Metabolic 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
- metabolic 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 Resources and Contacts, Chapter 17).

Explain that your baby has a metabolic disorder so they will understand why you need advice about expressing milk rather than advice on how to breastfeed.

Looking after Mum when breastfeeding

Taking care of yourself as a breastfeeding mum 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 is natural and this may cause a drop in your milk supply, but it won’t affect the quality of your milk.
- If you are expressing concentrate on expressing frequently to keep your supply, rather than focusing on how little you may be producing – your supply will adjust to your baby’s feeding demands.

- Babies with metabolic disorders often only need about half their feed as breast milk, so some drop in supply is no cause for concern.

What to do if you are unwell when you are breastfeeding

Contact your family doctor for advice if you 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 a metabolic disorder. Telephone the Metabolic doctor who will advise you about feeding your baby.

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 may help relieve pain. Antibiotics may also be needed.

What to do if you are unable to breastfeed

If you are unable to breastfeed because of illness seek advice from your Metabolic Team immediately by calling the 24-hour emergency number.

If you have to miss one or more breastfeeds

This is not unusual and you can:

- give expressed breast milk—if available from the freezer—after the usual amount of special formula
- if you don't have breast milk in the freezer, you can give an unlimited amount of special formula, whenever your baby is hungry
- ask your Metabolic Team about using a standard infant formula instead of breast milk
- keep in close contact by daily phone calls with your Metabolic Team until you return to your usual routine.

Stopping breastfeeding

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

In an emergency, follow the guidelines above and contact your Metabolic Team as soon as possible.

Cleaning and sterilising 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.

The boiling method

- Place the bottles, caps and teats in a large saucepan.
- Cover with water, making sure the bottles are full of water and completely covered. Cover the saucepan with a lid.
- Bring to the boil and boil for five minutes.
- Turn off the heat and allow to cool.
- Bottles and teats may be left in the covered saucepan until the next feed, or assembled and stored in a clean, dry place.
- 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.
- 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.
- Boil all equipment every 24 hours even if it has not been used in that time.

The 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.

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

Using sterilising solutions

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

- Follow the manufacturer's instructions when making up the solution, making sure the proportions of water and sterilising solution are accurate.
- After washing and cleaning the bottles, caps and teats place them into the diluted sterilising solution, making sure they are completely covered.
- Leave the feeding equipment in the solution for the time recommended by the manufacturer.
- 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.
- 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.
- Remake the liquid in the sterilising bath every 24 hours.
- Keep the solution away from children.

Chapter 9

Feeding your child: starting solid foods and beyond

Note: we've referred to babies as he in this chapter, but information applies equally to girls.

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, or siblings, 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 Metabolic 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 the way you introduce solids will be different to the way you would do it for a baby without a metabolic condition.

Counting protein

'When I went to buy another tin of formula once my baby was 6 months of age, I just assumed I should buy the follow on tin rather than the starter. But this is much higher in protein and we had to do some quick adjustments once the blood test results came back too high.'

As the variety of foods in your baby's diet increases you will learn about counting protein in their food. (See Counting Protein in 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 protein in foods list your dietitian will give you. Food will gradually replace some of the protein your baby is getting from breast milk or formula. If you are using a standard formula do not change to the over 6 months version (usually labelled 2 or 'follow on') without discussing this with your dietitian.

At first it's a good idea to write down the foods your baby has eaten—until you get used to the process of measuring and counting. Your dietitian will explain how to fill in food records, which may need to be taken to clinic visits or sent with blood samples. (see *Sam's Protein Planner* in Chapter 11)

The special formula or a supplement 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 best not 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 baby's mouth opens, place the tip of the spoon just inside. Babies take the food from the spoon in a sucking motion.

At first, baby may push the food out with his tongue. This doesn't mean baby doesn'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 but it is important to remember that all babies have their own ways and preferences where food is concerned.

Three steps to introducing solids

Solid food is introduced in three stages:

1. smooth, lump-free food
2. lumps and chunks that can be chewed
3. self feeding and finger foods.

Step 1: smooth, lump-free food

Fruits and vegetables are mostly low in protein, and make good first foods for your baby on a low protein diet. Smooth, lump-free foods—that is foods that have been sieved or pureed—can be made at home, or bought ready-made in jars or cans of commercial baby food. Strained fruits, fruit gels or vegetables are suitable.

Start with a small amount—one or two teaspoons. 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.

At this stage you will need to be counting protein in your baby's food, as you have shown by your dietitian:

- foods can be mashed, grated, diced and pureed more thickly than before
- the variety and amount of foods your baby eats can be increased – try mixtures of vegetables, fruit and baby cereal, low protein custard, low protein pasta with vegetables, (see Low protein recipes in Chapter 7 and other recipe books for ideas)
- sips of water from a sipper cup can be offered
- the special formula from a bottle or tiny amounts from a sipper cup need to be included.

MEAL PLANNING

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

- Aim to spread the number of grams of protein allowed throughout the day.
- Decide how many grams of protein your child can have for that meal or snack.
- Choose the foods you will offer.
- Choose one higher protein food such as cereal, potatoes, peas, or corn.
- Choose a lower protein vegetable or fruit such as carrots, green beans, peaches or pears.
- Measure the amount of food your child may eat at a meal to add up to the number of grams of protein available for that meal.
- If your child doesn't eat all the food offered, estimate how much is left and subtract it from the calculated grams of protein. He may want some extra later in the day.
- As your child eats more foods, he will need to consume less of his regular formula or breast milk so his total protein intake is appropriate for his needs.
- Be sure to let the dietitian know if your child is not eating all of the grams of protein allowed or regularly leaves some of their formula. The special or regular formula may need to be adjusted.

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

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 ten to twelve 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 so easy-to-clean baby equipment makes life easier.

If baby get used to sitting in a highchair or at a low table when eating, rather than sitting on your lap or the floor, it keeps him focused 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:

- low protein rusks or hard dry toast made from low protein bread (see Recipes in chapter 7)
- 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 practise with and one for you.
- Offer soft cooked table foods cut into small pieces at ten to twelve 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.
- Always stay in the room with your baby when he is eating finger foods or self-feeding.
- Babies can choke on large pieces of hard foods. If it is possible for a hard piece to be broken off, grate, cook or mash the food, foods such as raw carrot, celery or apple are best treated this way.

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
- offer vegetables and/or fruit at most meals and snacks
- 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 he is eating
- stay positive about food and formula and avoid power struggles.

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

Mealtime, family time

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.

Your child will want to have foods he can't eat. You will need to tell your child he is special and show him which foods he can have. The Metabolic Team will provide tips on how to avoid or manage mealtime problems with siblings and extended family.

The special formula

As your baby eats more solid food and less breast milk or infant formula, there will be changes with the special formula (called the 'supplement' after your child is twelve months of age). The supplement is still essential, but your dietitian will change the recipe, and the type of supplement (read more about the special supplements in chapter 5).

The special 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'* foods. These will depend on how restricted your child's diet is but for many children on low protein diets *'Yes'* foods are free and very low protein, such as apple, carrots and low protein foods. *'Maybe'* foods must be measured accurately, such as crackers, rice, pasta, cereal, potatoes and some vegetables. *'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. By this age you can also offer small pieces of suitable soft-cooked low protein family foods, there is still a risk of choking on hard pieces of food particularly if children run, play, laugh or cry while eating.

Your child will also be able to cope with set meal and snack times. You will also have become used to monitoring your child's protein intake and you will need to continue to count the protein carefully. To make it easier to do this:

- offer a set number of grams of protein at each meal and snack,
- continue to keep a record of grams of protein – food records may need to be sent in with blood tests and brought to clinic visits
- start introducing more low protein foods.

At this age you can give most fluids by cup. The special formula may be the only formula your child is having at this stage and your child needs to have their special formula/supplement every day in the quantities prescribed.

If your child is still drinking standard infant formula, ask your dietitian if cow’s milk may be substituted.

Make eating safe for young children and reduce the risk of choking by:

- not giving foods that can break off into hard pieces
- avoiding foods such as raw carrot, celery sticks and apple pieces—hard foods should be grated, cooked or mashed
- not giving hard lollies, corn chips or other small hard foods to young children
- staying with young children and watching them while they are eating
- having them sit quietly while eating
- never forcing young children to eat.

Your Metabolic Team can give you further advice.

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.

Your child will let you know when he’s 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. Allow him to ask for more if he is still hungry.

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

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

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.

Seven tips for reducing tension at mealtime

1. Respect that your child may not be hungry

After the age of twelve months children don't grow as quickly, which means their appetite won't be as large as it was in the first year.

2. Watch for growing independence

As toddlers start discovering that they are independent people, they may express their likes and dislikes more strongly. This is all part of growing up.

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 on a low protein diet. 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 grams of protein later, or the next day. Talk to your dietitian about ways to handle this situation and how much variation in the number of grams of protein eaten is likely to be OK.

It is never a good idea to force-feed a child. This often leads to fear of mealtimes and further refusal 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 he likes it. Praise your child for trying new foods.

4. Help your child understand their special diet

A child with a metabolic disorder may find it difficult to understand why he can't eat the same foods as others. As a result he may refuse to eat his 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 his need for a special diet. These explanations will become more complex as he grows older. Ask your Metabolic 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. By keeping the mealtime atmosphere relaxed this will be 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.

Children have short attention spans. Set aside twenty to thirty minutes for meals and ten to fifteen minutes for snacks. Seat your child at the table for meals when the food is ready. Forcing your child to sit for longer than he can handle may lead to him refusing food.

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

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

Allow your child to form his own opinions about his diet. Stay calm, and avoid showing your anxiety in front of your child—it 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 Metabolic Team is there to help if you are having trouble with feeding.

Chapter 10

Meal planning & healthy food choices

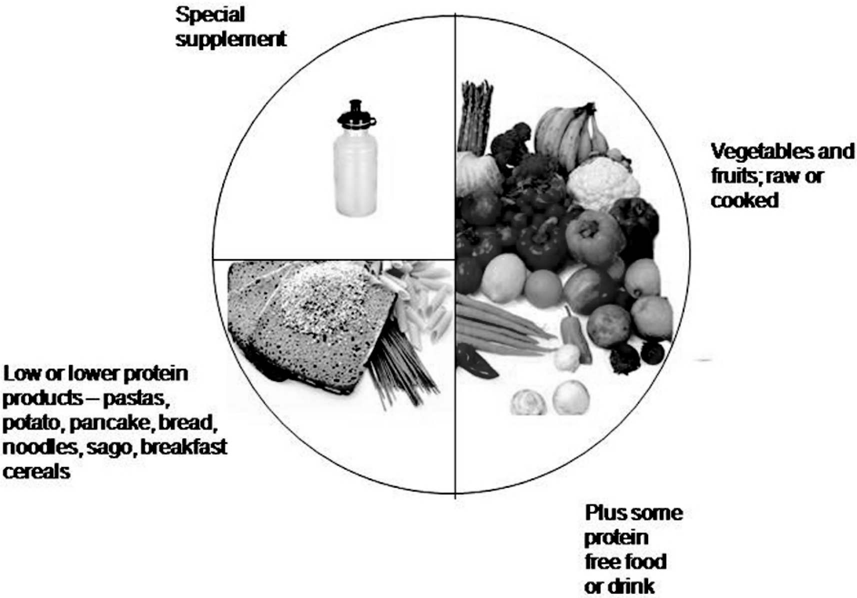
What makes up a low protein meal

THE FIVE PARTS OF THE LOW PROTEIN DIET

- Special formula or supplement (see Chapter 5)
- Counted protein from food (see Chapter 6)
- Low protein foods (see Chapter 7)
- Meal planning and healthy food choices (see below)
- Changes needed during illness (see Chapter 3)

Family meal times provide a good opportunity to talk about the day and also for children to learn about healthy food choices. From the time your child is eating food it is important to start incorporating a child's low protein food options into the family meal setting. This helps you to get used to doing it and helps your child to feel part of normal everyday family life.

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



What makes up a low protein meal?
Some people on a low protein diet may also be able to include small measured amounts of meat, fish, egg, chicken, dairy, nuts or legumes

Preparing a low protein meal & family meal at same time

Family food	**Low protein meal** *The following meal suggestions use low protein alternatives for bread, pasta, rice, flour, sauces and biscuits. If ordinary types are allowed, substitute these*
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 ▪ 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, mash or chips and salad	Sandwich mashed herbed vegetables between low protein cracker biscuits pre-cut into finger-sized pieces, tomato sauce, mashed or chipped parsnip OR roll mashed vegetables into

Family food	**Low protein meal** *The following meal suggestions use low protein alternatives for bread, pasta, rice, flour, sauces and biscuits. If ordinary types are allowed, substitute these*
	sausage shape using cornflour or low protein flour mix to bind then fry in oil & serve with tomato sauce, chips and salad
Beef patties in hamburger bun	Vegetable patty in low protein toasted bread or roll
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
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 crackers 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

Desserts are good "fillers" and often low in protein. You can use low protein breads and cereals in the dessert if these were not part of the main low protein meal:

- apple (or other fruit) crumble—make topping from low protein crumbed sweet biscuits or low protein flour, sugar and margarine and baking powder
- low protein pancakes with fruit, honey or sugar, lemon juice or cinnamon
- low protein rice or pasta with sugar or honey and margarine, cream, rice milk, Duocal™ or Calogen™
- low protein rice with fruit and cream
- sago or tapioca with various flavours.

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.'

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

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

Low protein meal ideas

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

Mediterranean and Asian meals

Many of the foods from these cuisines are easily adapted to a low protein diet. For example you can serve low protein spaghetti or rice with:

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

‘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.’

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.

Roasts, burgers, barbecues and more

Though someone on a low protein diet cannot eat meat in the same way that others do there are many ways a meal can be adapted so that they do not feel excluded from family foods. 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, parsnip, 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 low protein 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, put back in shell and reheat.
- **potato or sweet potato**—slice, layer and bake with sour cream
- **vegetables**– partly cook until crunchy, 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

'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.'

- **sweet and sour vegetables –** partly cook and serve hot with a sweet and sour sauce
- **low protein jaffles or toastie pies**—fill with mashed vegetables for a quick and easy meal
- **mildly curried vegetables**—add a curry paste or powder and cook with vegetable stock and serve with low protein rice
- **risotto style rice**—use diced vegetables, vegetable stock and flavourings to make a risotto
- **barbecue**—use a combination of partly cooked vegetables and fruits on a skewer and serve with a cold low protein pasta salad.

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. You can replace the usual high protein cheese sauce with a white sauce made with cream and cornflour or use one of the packet sauce mixes from the supermarket that are low in protein.

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. Lunch can be based on bread or crackers, with salads, fruit and other snacks.

Bread

- Low protein bread, rolls or flat bread
- Low protein crispbreads
- Rice cake
- Cold pizza slice
- Savoury low protein muffins or pikelets

Low protein sandwich fillings

- Avocado
- Grated carrot with sultanas
- Shredded lettuce
- Tomato and cucumber
- Beetroot
- Gherkin relish
- Olive tapenade (paste)
- Chargrilled vegetables
- Cold low protein vegeburger
- Honey
- Jam
- Scrape of Vegemite™ or alternative
- Scrape of cream cheese (check protein content)
- Low protein cheese and pickle

Fruit

- Fresh fruit (cut up and put into sealed plastic bags or small containers)
- Individual fruit snack packs
- Dried fruit

Salad

- cucumber, celery, capsicum and carrot sticks with a low protein dressing
- cherry tomatoes,
- mixed lettuce and salad leaves, with sprouts, spring onions and radishes

- stuffed olives, dill pickle, pickled or balsamic onions
- char grilled vegetables
- lightly cooked cold cauliflower or broccoli 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;
- low protein grated cheese, halved small pickled onions and grapes with a low protein or French dressing
- low protein pasta salad

Other lunch ideas

- different kinds of vegetable soup in a thermos
- low protein muffins, sweet biscuits, cake, fruit slice, fruit bars

Packed meals for PKUs, published by UK support group, The National Society for Phenylketonuria, has some great ideas for school and work low protein lunches that are suitable for a low protein diet for a metabolic condition. You can read it on the web at http://www.nspku.org/Documents/Packed%20Lunches.pdf.

Ask your dietitian about any products mentioned that you are not familiar with.

Snacks

These snack suggestions are ideal for after school, between meals, or any time:

- tomato salsa with low protein crackers or toast
- fresh, dried or canned fruit
- frozen fruit pieces e.g. grapes, rock melon, orange, banana, pawpaw, kiwifruit, peach or apricot
- vegetables with a salsa dip

- popcorn
- vege chips
- fruit ice blocks and icy poles
- Duocal™, Calogen™, 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 or jelly cup
- low protein pasta made into ‘cup-a-soup’
- low protein snack pot
- potato gems
- low protein breakfast cereal with rice milk.

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

Food on the run

Eating well on the run is all about being 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 7 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. There are tips for eating out in Chapter 13. 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.

Making healthy food choices

Many of the healthy food guidelines for Australia and New Zealand are important for a low protein diet. These include:

- eating plenty of vegetables and fruit
- choosing healthy fats
- controlling weight gain—some children and adults with metabolic disorders may become overweight, others may have trouble putting on enough weight.

- drinking plenty of water and consuming only moderate amounts of sugar and foods containing added sugar. This is important for healthy teeth.

Eat plenty of vegetables and fruit

Healthy eating guidelines for Australia and New Zealand recommend at least five serves of a variety of vegetables and fruit a day. Vegetables and fruit are quite low in protein and provide many important nutrients.

Choosing healthy fats

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 low protein diet. Food labels give information about the amount and type of fat the food contains. Choose from 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 low protein 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 hot chips, commercial biscuits, snack foods, cakes and pastries, cooking margarine, butter or cream. Some of these foods also contain **trans fats** which have a similar effect to saturated fats

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. A low protein diet tends to be low in omega-3 fats since fish, seafood, meat, 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 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 a low protein 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 a low protein diet

These tips will help you to lose weight:

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, which is mainly protein, as well as body fat. This will increase your blood levels of unwanted metabolites. In some metabolic conditions this could make you very ill. Gradual changes lead to gradual weight loss, which is more likely to be sustainable than a quick fix.

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 keep the metabolic condition in control while you lose weight, and may also increase the amount of protein you can tolerate in food.

3. **Supplement.** Take the amount of supplement your dietitian recommends. If you don't take enough you're more likely to break down muscle, which will cause blood levels of harmful metabolites 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 special supplement is best when you're trying to lose weight as some are lower in energy.

4. **Have your usual protein intake.**

5. Eat often and eat three meals and snacks if you need them 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 **Fat Facts** in this chapter for information on the best types of fat to choose. Reduce fat by:

- spreading margarine or butter thinly on bread or biscuits, or going without
- using only a little margarine or oil when cooking – about a teaspoon per serve
- adding herbs, spices, lemon juice or vinegar for flavour rather than using fat
- using low-protein low-fat salad dressing
- choosing low fat snacks such as fruit, vegetables, vegetable soups, sandwiches and salads rather than hot chips
- only eating high fat foods occasionally and in small amounts.

Keep these high fat foods to a minimum:

- hot chips
- cream and sour cream
- chocolate (including low protein)
- 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, but people with tyrosinaemia must avoid artificial sweeteners containing aspartame (see Chapter 3)

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 rather than juices, soft drinks and cordial.

9. Avoid fad diets particularly high protein diets or very restricted diets, which are unsuitable for someone with a metabolic condition. They may:

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

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 metabolic doctor, avoid them.

Gaining weight on a low protein diet

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. **Increase your daily supplement.** You will need a little more of your special supplement than you normally need to simply maintain your weight. Take it throughout the day in three or more lots.
2. **Maintain your protein levels.** Have the amount of protein recommended by your dietitian.
3. **Eat often.** Eat at least three meals a day and three snacks.
4. **Fill up on low protein foods** such as low protein pasta and rice, low protein bread, low protein biscuits and low protein cereals.
5. **Choose 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 low protein diet’** earlier in this chapter for a list of foods that are high in fat and sugar.)
6. **Try energy supplements.** If you only have a small appetite supplements can be useful. You can try:
    - a fat and carbohydrate supplement such as Duocal™ (available on prescription)
    - carbohydrate supplements such as Polyjoule™, Polycose™ or Polycal™
    - a fat supplement such as Calogen™

      These can be added to your protein supplement, used on cereal, as a shake or in low protein custard. Polyjoule™, Polycose™ or Polycal™ can also be added to cordial or fruit juice.
7. **Drink energy drinks.** Instead of drinking water, tea or coffee, drink soft drinks, sweetened fruit juice or a low protein shake.

Dental care

Tooth decay and dental erosion can affect the teeth of any child. Children on a low protein diet are at greater risk, and adults also need to take good care of their teeth.

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

There is a greater risk of dental decay and erosion for children and adults on a low protein diet because:

- **Sugary foods and acidic drinks may be consumed more often**. The diet for children on a low protein diet includes less protein from foods than usual, 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.

Many special supplements are both acidic and sweetened. They are usually given as a drink, gel or paste several times a day. The 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.*

Dental disease can be prevented by:

- toughening the teeth with fluoride
- giving the teeth a rest from food and drinks
- having regular check-ups with a child-friendly dental practitioner.

Toughening the teeth with fluoride

Fluoride makes teeth tougher and more resistant to decay. You can ensure that your child's or your own teeth get access to fluoride by:

- Using toothpaste to get fluoride onto the teeth—start using fluoride toothpaste as soon as the teeth appear.

- If you live in an area that has fluoride in its water supply (such as a major city) use a low fluoride junior toothpaste for young children—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.
- Wiping just a smear of toothpaste onto your baby's teeth using a face flannel or gauze—it is important that an adult takes responsibility for applying this very small amount of paste to baby's teeth.
- 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 after breakfast and last thing at night (and avoid night-time sweet drinks or snacks afterwards).
- Adults should use a full-strength fluoride toothpaste twice a day—before bed and after breakfast.

Give the teeth a rest from food and drinks:

- Only put infant formula, expressed breastmilk, special 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.
- Aim to move to a feeder cup by the age of twelve months.
- Encourage your child to 'drink up' rather than sip a drink slowly.
- Aim for three meals a day and only two or three snacks between meals, rather than continual snacking. Snacking on lollies and biscuits increases the risk of decay.
- Avoid giving sweet food or drink at bedtime.
- Water is the best drink to have apart from the supplement— offer water after each supplement drink and often during the day.
- Keep fizzy drinks, cordials and juice for mealtimes, special occasions or illness.

SUGAR OR SUGAR-FREE?

Sugar is useful in a low protein diet as it helps to provide energy. Sugar-free foods don't do this – discuss with your dietitian whether they are appropriate for you or your child. Artificial sweeteners containing aspartame are also not suitable if you or your child have tyrosinaemia. When eating sugary food it is important to avoid constant snacking or sipping, to give teeth a rest, and to maintain good dental care.

Go for regular check-ups

Begin in your child's first year year by taking him to the dentist with you when you go—and staying with him while the dentist sees him. Your dental practitioner will be able to pick up signs of decay and erosion much earlier than you or your doctor. You can:

- Find a child-friendly dental practitioner who will help your child get used to regular visits—this can make it a lot easier as they get older.
- Be sure the dental practitioner knows that your child (or you) have a metabolic condition and understands what it is, and the diet needed to treat it.
- 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.

Chapter 11

Encouraging independence:

Talking about the metabolic disorder with your child

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 blood levels, and explaining the metabolic disorder to other people. As they grow up, it is important to start sharing this responsibility with your child.

Why encouraging independence is important

Fostering independence in managing their condition has a number of advantages including:

- encouraging **acceptance**
- helping them develop a good **understanding** of their condition
- increasing their **confidence** in controlling their condition
- helping you and your child to **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 that your child will be less likely to blame you for his or her treatment.

When children with metabolic disorders 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 responsibility for their condition.

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

What to expect of your child at different stages

Toddlers: two to five years

At this age most children:

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

Early childhood: five to eight years

Children in this age mostly:

- understand that they have a special condition
- understand that they can’t eat high protein foods
- know they have to have medicines and special drinks if they are sick and make some choices as to how these are taken
- have a basic knowledge of why the special supplement or medication is important
- help you prepare the special supplement
- practise using scales to weigh foods and count protein
- have ideas about meal choices
- learn to select low protein foods in social situations
- can assist in taking their own blood samples.

Late childhood: eight to twelve years

By the time they reach the pre-teen years most children:

- are able to prepare their own special supplement

- can weigh foods accurately and count the grams of protein
- know they have to have medicines and special drinks when they are sick and monitor their intake of these
- are able to make appropriate meal choices
- are able to take their own blood samples if collected on a card, or visit the pathology service or laboratory themselves
- have a basic understanding of their condition and the low protein diet
- know how frequently they need to have blood tests
- understand that the special supplement helps provide nutrition and helps keep their metabolic condition in control
- can read and calculate the grams of protein from product labels with assistance.

Early teens: twelve to fourteen years

Young teenagers have a greater knowledge and:

- know what their sick day needs are and how to contact the hospital if necessary if they are unwell (this is essential for some metabolic disorders but not others – see Chapter 3)
- prepare their own special supplement all the time
- take their special supplements and medications as prescribed
- know what foods to count for the low protein diet
- can read and calculate the grams of protein from product labels without assistance
- are able to make appropriate meal choices
- can prepare some basic meals for themselves
- are able to take their own blood samples if collected on cards paper, or visit the pathology service or laboratory themselves
- have an understanding of what causes their condition, enzymes, amino acids and the effects of illness
- know the healthy range for their blood test results
- understand risks and effects of high blood levels of harmful metabolites
- make an effort to expand low protein diet choices
- are able to keep a food diary to record protein intake
- attend an adolescent metabolic clinic or have a plan in place for transition to an adult metabolic service.

Late teens: fourteen to eighteen years

By now your child is capable of taking responsibility for most of his or her care. They:

- know what their sick day needs are and how to contact the hospital if necessary if they are unwell (this is essential for some metabolic disorders but not others -see chap 3)
- prepare their own special supplement
- take their special supplements and medications as prescribed
- know what foods to count for the low protein diet
- can read and calculate the grams of protein from product labels without assistance
- are able to make appropriate meal choices
- can prepare low protein meals and other family meals
- remember to take their own blood samples and send blood samples for testing if collected on cards, or visit the pathology service or laboratory themselves.
- are able to make dietary changes suggested as a result of blood results
- have a thorough understanding of their condition and its effect on the body
- can explain how their condition 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 metabolic doctor, dietitian or nurse when required
- are aware of the pregnancy risks associated with their condition if appropriate (see Chapter 3)
- attend an adolescent metabolic clinic or have a plan in place for transition to an adult metabolic service.

Talking about the metabolic disorder with your child

Talking to your child about their condition and its day-to-day management is beneficial even at an early age. The Metabolic 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.

'Mum always used to say to me if you eat the wrong foods you won't grow properly or be able to run so fast—it's up to you. 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 and good at football.'

If their metabolic condition is one in which they can get very sick when they have the usual childhood illnesses or accidents, they need to understand why they have to drink their sick day drinks and go to the hospital. They need to tell you or ask school to ring you when they feel unwell.

Your child will be asked questions about their diet and condition. 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?
- why did you have to go to the hospital when you had the flu?

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 my condition; 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.
- I'm used to having the supplement. It's like medicine – if you need to take it to be healthy, you take it.
- Sometimes I can get really sick if I have the flu – worse than usual.

I’m OK now.

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

Games that help your child to understand

Regular discussions about their condition with your child will help your child to feel more comfortable talking about it, and give them the confidence to ask you questions and discuss their concerns. You can help your child to understand their condition 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 scrapbook
- letting your child rate new foods or meals from one to five to encourage them to try new foods
- putting their blood results on a wall chart so they can monitor their progress
- cooking low protein recipes to take to school or share with the family
- trying different ways to take their sick day drinks when they are well and rating them from one to five
- a star chart/reward system for taking blood tests, drinking their supplement and trying new foods.

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

Daily protein planner

It is more fun for your child and you will be assured that you know what is happening if you count the daily grams of protein 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 gram, place a magnet on the board. As the grams of protein are eaten during the day, your child removes the magnets from the board until they are all gone. Another option is to laminate a chart and stick it to the fridge. Write on it with a whiteboard marker and keep a tally that way.

Sam's Protein Planner	**grams**
Protein for the day: ***11***	
Breakfast:	*3*
Snack:	*1*
Lunch:	
Snack:	
Dinner:	

Chapter 12

Preparing for childcare, preschool and school

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. If your child has a metabolic condition which means they can get very sick from the usual childhood illnesses, then you are even more likely to be concerned.

With preparation children with metabolic conditions can make this transition and keep their condition well controlled. There needs to be ongoing support and communication between the family, school and the Metabolic Team.

Each child care facility and school is run differently. The following suggestions are a guide for planning and discussing your child’s metabolic 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.

Discussing the metabolic condition with carers and teachers

Who you need to inform

It is important that teachers and others at child care, preschool or school understand why your child needs a special diet, and why the diet needs careful supervision, and also that they know about particular concerns with illness.

'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 and adjust her diet around it. I find talking to her about what she can and can't have is the best. She's almost four now and understands pretty well.'

You should inform the following people about your child's metabolic condition:

- the principal, admissions staff, and preschool supervisor (when you're planning your child's enrolment)
- the class teacher
- the school nurse—for support— particularly if your child is unwell and as an education resource
- the supervisor of after school care
- the tuckshop or canteen supervisor or cook.

The school needs to be reassured that, apart from needing a special or restricted diet, and in some conditions, special care during illness, your child is perfectly healthy.

What to discuss with staff

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

Anyone caring for your child needs to know that:

- Your child's condition is an inherited, non-contagious condition.
- Children with this condition cannot break down protein in foods as well as most of us do.
- All children need a certain amount of protein for growth and repair of the body, but in your child's condition extra protein can damage them.

'When telling teachers your child has a metabolic condition, you're very aware that you don't want to label your child as potentially having brain damage or being 'sick'. So I quickly tell them she is normal like everybody else, because she's eaten the right food.*'*

- Staying on a protein restricted diet keeps the metabolic condition in control so that the child will develop as well as possible.
- Eating the wrong foods will not make them immediately sick (like a severe allergy), but will have a detrimental effect over the day or longer term.
- Your child is on a very specific diet that is calculated by a specialist team, with the amounts of foods 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.
- You must be informed if your child has eaten food that is not allowed, or does not eat foods that are sent from home.

If your child has a metabolic condition requiring special treatment during illness, the school must inform you if your child is unwell or has an accident as prompt treatment may be needed to prevent more serious problems occurring.

At the end of Chapter 2, you'll find sample letters which offer a simple explanation of your child's metabolic condition 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 Metabolic Team can answer questions and review the main points.

'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.'

Food at school

Each child care 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 are 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 special supplement to school

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

Lunches, parties or 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 your child can share food that you supply, such as low protein pikelets, or provide the recipe and ingredients, (See Chapter 13 for more ideas about food for special occasions.)

'I arranged with the teacher to keep some low protein cakes in the staff freezer for the morning when the birthday child brings in their cake. It defrosts by recess when they are ready to celebrate.'

'One day I gave my daughter my sandwich in her lunch box instead of hers. When I went to eat mine I knew her lunch break was over. I was in such a panic. But she had told her teacher and he had bought her something from the canteen that she could eat. I was so proud of her.'

School camps or overnight excursions require more planning (see School Camps and Excursions in chapter 13).

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:

- knowing which foods are OK and which to avoid
- bringing home uneaten food in the lunch box so that you can calculate grams of protein
- buying only low protein food from the canteen
- 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 because of the diet.

Children often find it difficult being different from other children, and they may 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 children who do not have a metabolic condition)
- 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 13

Parties, celebrations, camps, school excursions, eating out

Children's birthday parties

It is normal for parents to worry that their child will overeat at a party but for parents of children on a low protein diet this can be a big concern. Here are some tips for managing your concerns:

- to help minimise the risk of overeating, make sure your child doesn't miss any meals or snacks before the party
- give fewer grams of protein at the meal before the party to leave a little room for party extras
- avoid major discussions about food in front of your child and their friends
- be realistic, and expect that your child may have more protein than usual and cut back at the next meal.

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 low protein foods. Older children may be more aware of the differences between low protein and regular foods. Here are some tips to help make the day a success:

'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 vegie chips.

- include lots of non-food fun activities
- 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 protein for everyone—include some of your child's favourite low protein 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.

Making a low protein birthday cake

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

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

- Use a favourite low protein cake recipe and decorate it with icing and low protein lollies—try cutting the cake to make simple shapes before icing it.
- Make a low protein ice-cream or gelato 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 low protein 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.

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. Low protein lollies can be used as prizes and for the lolly bags, also small packets of chips (including vegie chips) or biscuits.

'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.'

Games and activities

These ideas can help take the focus off foods:

- Arrange for a clown, a special story teller or a face painter to visit
- Hold the party in a park with picnic food for activities such as kite flying, Frisbee throwing, a game of cricket or a treasure hunt for small toys or party favours instead of lollies.
- 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.
- There are plenty of games that don't involve food—old-fashioned games such as Red Rover, What's the Time Mr. Wolf? Musical chairs, Pass the parcel. Your local library has plenty of books about children's party games if you find it difficult to think of some.

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 low protein chocolate, low protein 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 Low Protein Party Food Ideas section on page 5, highlighting the foods that are easy to prepare and that other children at the party will also enjoy.

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

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 low protein 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.
- Let the host know that your child should not have milk shakes or flavoured milk—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 extra but check with the restaurant that this is okay first.

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 the child who has a metabolic condition. Most children of this age can identify high and low protein foods and it is a matter of parents trusting them. If they've shown they can do this at home, they're likely to carry it through when they are out socially.

'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.'

'I buy Natural' carob and milk-free Easter eggs that 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.'

Low protein party food ideas

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

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

Drinks

- soft drinks and cordials
- fruit juice cocktails
- use fun straws and cups

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

Lollies

Write a list of your favourite low protein lollies here	

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.

At Christmas time, bake some low protein mince pies and low protein Christmas pudding or cake. Encourage relatives to give low protein chocolate, toys or non-food treats.

At Easter time you can make low protein Easter eggs by filling plastic casings from craft stores, such as Spotlight™, with melted low protein chocolate. Try melting low protein chocolate and a small quantity of dark chocolate together and putting them into different moulds. Shop around for some of the cheaper chocolate eggs that are lower in protein.

Camps and excursions

'She was seven when she went on a four-day school camp. I sent a book so her teachers would understand her diet. She added up the grams of protein and I checked it when she got home.

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 on a low protein diet about how to prepare for a camping trip, going away with friends or staying the night at a friend's place, see the Quiz in Chapter 18.

Getting organised

Here are some tips to help you prepare for an overnight or longer camp:

- discuss your requirements with the camp supervisor, including the need for special supplement and any special care needed for illness or accidents
- try to obtain copies of the menus so you can help plan your menu for the camp
- pre-weigh and package the special supplement into separate bags so that it just needs to have water added at the camp
- take a supply of low protein bread, pasta, crackers, biscuits and low protein chocolate to keep your energy levels up
- include some free foods for midnight snacks
- give the camp supervisor a clear meal plan
- keep your mobile phone switched on in case there are problems.

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
- vegie chips – pack in a plastic container so they don't get crushed

'I've always had a meeting with the teacher well before the camp. However last time the teacher in charge of her diet wasn't there at meals so she missed out and had to fill up on the extra supplies she had taken. Next time I will get her to ring me at meal times to be on the safe side.'

- cherry tomatoes
- rice cakes or corn thins
- lollies (without milk or chocolate) or low protein chocolate.

Teenage and adult parties

Eating low protein foods in the time leading up to the party should give you enough protein to enjoy the party without going overboard. There will be times when you go over your intake of protein. Just don't overdo it and make sure you eat less protein for the next day or two and lots of low protein foods.

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, knowing 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
- vegie chips

Eating out

There is no reason why you can't enjoy eating out with friends and family. There are many dishes you can enjoy once you know what is suitable.

Takeaway

Look for these foods:

- hot chips*
- apple pie*
- fruit salad
- salad
- soft drink
- juices
- mashed potato* and gravy
- hash browns*
- corn
- sushi (no fish)*
- salad roll*
- 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 or give nutritional content of foods on their website so you can work out the protein content. Note however that 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.*

Cafés

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 curries or stir fry
- vegetable based risotto or noodles*
- 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 but with no cheese – 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
- meringue or pavlova* with fruit and cream

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, chapattis, pappadums, parathas*
- dishes made with sago
- vegetable dishes and curries 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 eat low protein versions. Eat a small serve and be cautious about protein intake for the rest of the day.*

Cooking meals for family and friends

Adapting low protein meals for a higher protein intake 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.

Very low protein meals are not suitable as a main meal for someone not on a low protein diet.

Chapter 14

Low protein diet: teenagers & adults

Staying on the diet

A low protein diet for most metabolic conditions is for life. It may become a little more flexible as you get older, once you have stopped growing. But for many metabolic conditions there is a risk of serious consequences if you overdo your protein intake.

> If you want to broaden your diet or stop your supplement then it is essential—an absolute *must*—to talk to your doctor and dietitian. They will understand why you may want to do this. It is important that you understand the effects of not strictly following your diet and treatment. Any changes must be done in the best possible way with the least amount of side effects and damage to your health. Your doctor and dietitian will work with you to help fit your diet and treatment into your lifestyle.

If you have been attending a Metabolic Clinic since you were very young, it is important that **you** understand about your metabolic condition and its management. You are now the one responsible for looking after yourself. Read up about your particular metabolic condition in Chapters 1 to 3 and ask your Metabolic Team to explain anything you do not understand.

If you have not been to clinic recently do not be embarrassed about getting back in touch. The team will be delighted to see you again and bring you up to date with the management and treatment of your metabolic condition.

Important!
If you have a metabolic disorder that can cause you to get very sick if you get ill (for example catch the flu), have an accident or need an operation, you **must** understand clearly the changes needed to your diet and medications. See Chapter 3.

Your low protein diet

The diet for many teenagers and adults with metabolic disorders can often be more flexible. You need to discuss this with your Metabolic Team.

Teenagers and adults may be able to tolerate more protein in their diet, and to eat more normal (i.e. not low protein) foods—such as bread, breakfast cereals, rice, corn and potatoes. Discuss what **you** can tolerate with your doctor and dietitian.

Many resources are available to help make staying on your 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.

Guidelines for the low protein diet for teenagers and adults

- **Take your supplement**—if you take a special protein supplement make sure you take the full amount every day and have it two to three times a day with meals so it's spread throughout the day (ideally over 12 hours). If you take energy, vitamin or mineral supplements take these daily as recommended by your Metabolic Team. These supplements are important to the control of your metabolic condition and for your health and wellbeing.
- **Count protein**—do this as directed by your Metabolic Team (see Chapter 6), and eat some protein three times a day at meals. Don't save your protein allowance up for one meal.

- **Understand any changes needed when you're sick**—see Chapter 3.
- Attend the Metabolic Clinic at least once a year so that the team can update your treatment and your diet can be checked to make sure it is adequate.
- **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 protein.
- **Drink plenty of water**
- Eat the **right sorts of fats** and limit your overall fat intake (see choosing healthy fats, 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**.
- Know how alcohol affects your metabolic condition. If you choose to drink alcohol, **drink it in moderation** and avoid those with a high protein content e.g. cream liqueurs and Advocaat™.
- Take care to store and cook food safely so that it retains its **freshness and nutrients**—be aware of its use before or use by date

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 a metabolic condition 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. Your low protein diet and/or special protein supplement should provide your body with enough protein. Discuss this with your dietitian if you’re concerned.

Commercial ‘protein and amino acid powders’ that promise to increase muscle bulk are **not** appropriate for a person on a low protein diet as they will increase your blood levels of harmful metabolites. Your diet and/or special protein supplement provide the protein your body needs and helps keep your metabolic condition 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 health and fitness and accepting the metabolic condition 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 it 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.

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 tell us that only five per cent of women are genetically programmed to look like today's female models—that leaves 95 per cent—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, practise a skill in sport or the arts or read books.
- **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.

Alcohol

Discuss with your Metabolic doctor how alcohol can affect your metabolic condition. Everyone needs to know about responsible drinking but having a metabolic condition does not necessarily stop you from drinking moderate amounts of alcohol. Keep in mind the following:

- protein in alcoholic drinks needs to be counted in the usual way
- 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.

The legal drinking age in Australia and New Zealand is eighteen and sensible drinking guidelines include the following:

- men—no more than four standard drinks per day
- women—no more than two standard drinks per day
- have two alcohol-free days a week
- excess alcohol can contribute to overweight so you need to drink less than these amounts to lose weight.
- 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.

A standard drink may be less than you think

1 standard drink	Volume	Protein
1 small glass wine	100 ml	0.2g
1 nip spirits or liqueur	30ml	depends on ingredients
1 glass regular beer	285 ml	0.9 g
1 can light beer	375 ml	0.8 g
¾ 300ml bottle alcoholic soda	260 ml	depends on ingredients

Support groups

Support groups and networks can make a big difference in the lives of people with metabolic conditions and their families. In Chapter 17 you'll find contact details for local support groups, and a number of internet sites that may be helpful.

Pregnancy

An increasing number of women with metabolic conditions are having successful pregnancies. For most metabolic conditions affecting protein metabolism, there is no risk to the baby if you have a metabolic condition as long as you are well and healthy. In Phenylketonuria (PKU) high blood levels are a risk to the baby's development but in other metabolic conditions this does not seem to be a problem.

Pregnancy will affect your own metabolic control requiring changes in your treatment, blood test monitoring and diet during the pregnancy and sometimes immediately after the birth. When you are planning a pregnancy talk to your Metabolic Team about the issues for your particular condition and the risk of your baby having your metabolic condition (see Chapter 3 and for an explanation of genetics see Chapter 1).

Remember it is important to:

- **Be healthy before pregnancy**—have your blood levels in good control and try to be in a healthy weight range.
- Check with your Metabolic dietitian that your intake of **vitamins and minerals** is adequate before you get pregnant as well as during the pregnancy. In Australia and New Zealand all pregnant women and women planning pregnancy are advised to take folic acid, one of the B vitamins. You may or may not need an additional supplement.
- **Maintain close contact with the Metabolic Team** before and during pregnancy. Let your clinic know as soon as you know you are pregnant, you may need more regular blood tests. Changes to your diet, supplements and medications will be necessary as your pregnancy progresses. You will need more protein in your diet as your baby grows and you may need to eat some foods you normally exclude to achieve this. The Metabolic Team will work

with your obstetrician (pregnancy doctor) to provide the best possible care for you and your baby.

- **Get an update of your sick day management plan and follow it carefully during any illness** and notify your Metabolic Team of any problems you are having such as morning sickness, vomiting or nausea.

Your diet before or during pregnancy will not alter whether or not your baby will have your metabolic condition. That will be coded in the genes and can't be changed.

The first 13 weeks

The amount of protein you can eat may not change much during this time. During the early part of your pregnancy, you may feel tired and a little sick. It is important to maintain your usual food intake but you may need to change when you eat. The following may help if you're feeling nauseous with morning sickness:

- small amounts of food very often – eating every fifteen to thirty 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
- have lots of water to drink but it may be best in small frequent amounts
- biscuits, bread, fruit, pasta and cereals help settle your stomach
- drink cordial, sports drinks, ginger ale or cola or eat ice and icy poles—if you become dehydrated you will feel worse
- having something to drink and eat while still in bed may reduce nausea once you get up in the morning
- having your supplement is important, try eating something to settle your stomach fifteen minutes before you start your supplement – a different flavour, making it more dilute and smaller amounts more often may help.
- it is important to eat enough to prevent weight loss, ask your dietitian for ideas if this is difficult.

If you have a metabolic disorder that can cause you to get very ill when sick or not eating properly, then you will need to be especially careful during this time in the pregnancy. Keep in close contact with your Metabolic Team.

Also during this time:

- your clinic will advise how frequently you need to have blood tests
- weight gain is supposed to be slow, but weight loss is not good for control of your metabolic condition or your baby's growth
- you may require a supplement of DHA (docosahexanoic acid), a special type of (omega-3) fat, as well as folic acid.

A healthy weight gain is 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 cause your blood levels of harmful metabolites to rise. From the time the baby starts to grow more rapidly, weight gain of up to half a kilogram each week is common. For most women, ten to thirteen kilograms is a healthy weight gain during pregnancy.

From 14 to 26 weeks

During this time:

- your blood test results may be lower than usual as the baby starts to grow much larger – you may be able to start eating more protein in foods—your dietitian will advise you
- expect weight gain of around half a kilogram each week.

From 27 to 40 weeks

During this time:

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

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 or energy drinks to make sure you are having enough energy (calories or kilojoules)
- if exercise is causing weight loss, you will need to reduce the amount or intensity for a while.

Obstetric care during the pregnancy

Your obstetric care before the birth will be the same as if you didn’t have a metabolic condition. The only difference will be that you are on a low protein diet and in more regular contact with your Metabolic Team. Your Metabolic Team will advise your obstetrician, local doctor or midwife about treatment for your metabolic condition during pregnancy. They will also advise on any special care needed around the time of the birth.

During and after birth

During and after the birth it may be necessary depending on your metabolic condition, to keep a close watch on blood levels of harmful metabolites as these can increase very suddenly, particularly immediately after the birth. Your Metabolic Team will advise your obstetrician if this is necessary and how best this can be managed.

Like all babies born in Australia and New Zealand, your baby will be tested for many different metabolic conditions and other problems soon after birth. Your Metabolic Team will advise you and the hospital when the test should be done and if any special care is needed for your baby.

Contact the hospital where you will be having the baby well before your due date, to plan for your diet during the admission. Your Metabolic dietitian can help plan your meals with the dietitian at the hospital.

Breastfeeding is ideal for babies and helps your weight (and shape) return to normal. Breastfeeding should be possible. You will most likely need to eat a protein and energy intake similar to your intake during the last few months of your pregnancy. Regular blood tests will check that your intake of protein and energy is adequate.

Chapter 15

Overseas travel

For people with metabolic conditions, holidays and travel away from home, particularly if 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 letter at the end of this Chapter).

The covering letter should list the name and quantity of all the special supplements, medications and foods 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 out of or into another country.

Carrying prescription 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 http://www.hic.gov.au.

Organising supplies for your trip

Always carry a little extra of medications, the special supplements and foods in case of delays or losses. For longer periods of travel, you will need additional scripts from your doctor so you can obtain extra supplies before you leave. Allow plenty of time for this to be organised.

Check with each airline you are travelling with on current restrictions as to what you are allowed to carry in hand luggage and plan around this.

Lost luggage

It’s a good idea to carry extra medications and 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.

Formula or supplement can be made during long flights by adding the measured powder to boiled water.

Precautions against illness

Traveller’s diarrhoea and other illnesses are more common when travelling. You will have more fun if you don’t get these. Reducing the risk of getting these illnesses is essential in some metabolic conditions (see Chapter 3)

Advice on healthy travelling is available at:

- http://www.smartraveller.gov.au/tips/travelwell.html
- http://www.welltogo.com.au/abcGuide/
- http://www.safetravel.govt.nz

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 metabolic conditions 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 metabolic conditions 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 are posting supplement (or formula) overseas include a copy of the doctor's letter and local contact numbers, if known, in the package.
- Ask your Metabolic Team to locate a Metabolic doctor or clinic you can contact when you're overseas in case you need assistance or run into problems.
- Take letters from the team with you explaining the metabolic condition, treatment and diet. See if you can get these translated for the countries you are going to. These can be given to the tour organizers if you are going on an organized tour. You will find this much easier if you use a travel agent.
- 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 out travel insurance for the trip
- Take a dictionary to help with translation

- Arrange to have someone at home on standby to post any items you may have forgotten or lost.
- Before you leave, you may want to search the web or ask your clinic for information about metabolic associations in the country you intend to visit.

Have you packed?

- medications
- formula or supplement
- mixing container
- scoop, tablespoon measures, if used
- vitamin and mineral supplements if needed
- prescriptions
- low protein foods
- customs letter
- letter explaining the metabolic condition
- scales, if needed
- diet information
- food for your trip

CUSTOMS DECLARATION

Dear Customs Officer,

RE ……………………………………………………………………….

Name….………………………..……………DOB……………………

Address…………………………………………………………………

Medical record no…………………………………………………….

………………….. has a very rare metabolic condition. 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 and is formulated as a white powder for reconstitution in water.

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 / New Zealand 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 medications (only available on prescription):

…………………………………………………………………………
…………………………………………………………………………
…………………………………………………………………………
…………………………………………………………………………

Yours sincerely,

Dr

Title

Chapter 16

Financial assistance

Special products

Special supplements for people with metabolic conditions are mostly available on prescription and are fully subsidised in New Zealand and Australia. You will pay the prescription cost.

New Zealand

In New Zealand, some special foods for people with metabolic conditions are also available on prescription (Loprofin all purpose baking mix, pasta spirals, spaghetti, macaroni and rice). Other foods can be purchased directly from the supplier (ask you dietitian for contact details/order forms).

Australia

Special low protein foods are not on prescription but can be purchased directly from the supplier. Contact details/order forms for these products are available from your metabolic dietitian or nurse.

Government allowances

New Zealand

Ask your doctor or social worker for advice on the financial support that may be available, contact Work and Income New Zealand (phone 0800 933 922, or visit http://www.workandincome.govt.nz).

The usual prescription charges apply when obtaining special 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 metabolic conditions 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 supplements on the lower prescription rate.

2. The Carer's Allowance

Parents of children with metabolic conditions 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 $98.50 per fortnight (Sept 2007).

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—this needs to be completed by your child's doctor.

3. The Inborn Errors of Metabolism Allowance

People with some metabolic conditions 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 a low protein diet for their metabolic condition, submitting regular blood tests where appropriate, and attending clinic. The IEM Allowance is currently paid at the rate of $217 per month (September 2007) to people of all ages and can be 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 Metabolic Clinic and need to be signed by your doctor.

Chapter 17

Resources and support groups

Australian support groups for metabolic conditions

MDDA Australia (Metabolic Dietary Disorders Association)
MDDA is a national association for people with a variety of metabolic disorders. It provides support, social activities, conferences and a quarterly newsletter. Contact:

MDDA, P.O. Box 33, Montrose, Victoria 3765
Tel: 1800 288 460 (Aust) +613 9728 8460 (outside Australia)
email: mdda_rebecca@iprimus.com.au
http://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 and some members with other metabolic conditions. It publishes a quarterly newsletter with information, updates and recipes, much of which is relevant to other conditions requiring a low protein diet. 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.

Current contact details and previous copies of their newsletter are available from the PKU Association of NSW, C/o Treasurer, 84 Undola Road, Helensburgh NSW 2508

http://www.pkunsw.org.au

PKU tree This is a web based support network for families with PKU and includes recipes.
http://www.pkutree.com.au/

New Zealand support groups

NZORD
This organisation for people and families with rare disorders covers some metabolic disorders.
http://www.nzord.org.nz

parent to parent NZ.
A support and information network for parents of children with special needs.
http://www.parent2parent.org.nz/start.htm

Breastfeeding support and information New Zealand

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

Plunketline
Free telephone support 24-hours day, seven days a week on 0800 933 922.
http://www.plunket.org.nz//AM/

La Leche League
Telephone help at any time. Local support groups meet regularly in some areas. Books and leaflets are available and the organisation has a free lending library.

Phone (04) 471 0690 for the name of your local support group leader (and for telephone advice), check your local White or Yellow Pages, or email lllnz@clear.net.nz
http://www.lalecheleague.org

Australia

Breastfeeding support and advice is available from your midwife, maternal and child health 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. (This organisation was formerly known as the 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
Toowoomba	(07) 4639 2401
South Australia and NT	(08) 8411 0050
Northern Territory counsellor contact	(08) 8411 0301
Tasmania	(03) 6223 2609
Tasmania—North	(03) 6331 2799
Victoria	(03) 9885 0653
Western Australia	(08) 9340 1200

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

Overseas associations, support groups and sources of information for metabolic conditions

See Chapter 7 for websites with recipes.

National Urea Cycle Disorders Foundation (USA)
http://www.nucdf.org

Screening, Technology And Research in Genetics (STAR-G) Project
http://www.newbornscreening.info/Parents/facts.html

Organic Acidemia Association, Inc. (USA)
http://www.oaanews.org

MSUD Family support group (USA)
http://www.msud-support.org/

New England Connection for PKU and Allied Disorders (USA)
http://www.necpad.org

Canadian Organization for Rare Diseases (CORD)
http://www.cord.ca

European Society For PKU and allied disorders treated as PKU
http://www.espku.org

Tyrosinemia—links and discussion group
http://groups.msn.com/tyrosinemia

Groupe Aide Aux Enfants Tyrosinemiques Du Quebec (GAETQ)
Hereditary Tyrosinemia Group (French)
http://www.cegep-chicoutimi.qc.ca/gaetq

International organization of Glutaric Acidemia
http://www.glutaricacidemia.org/

Climb. The National Information Centre for Metabolic Diseases (UK)
http://www.climb.org.uk/

National Organization for Rare Disorders (USA)
http://www.rarediseases.org

Association for Neuro-Metabolic Disorders
5223 Brookfield Lane, Sylvania, OH 43560, USA
Publishes a newsletter (3 times a year).

The Genetic Alliance (formerly the Alliance of Genetic Support Groups)
http://www.geneticalliance.org

UK PKU association
http://www.nspku.org

American PKU Associations
http://www.pkunews.org
http://www.pkuil.org
http://www.pkunetwork.org

Chapter 18

Quiz and problem solving

Use this quiz to test your knowledge or as a fun way of teaching your child about their condition. Have a go at answering the questions first, then look at the answers at the end of the quiz. People with metabolic disorders have provided many of these answers and ideas—some of which you may like to adopt for yourself.

Qu 1. How long should someone with a metabolic condition stay on the low protein diet?
Answer

Qu 2.What makes your blood levels of harmful metabolites go up?
Answer

Qu 3. When can you have more protein in your diet ?
Answer

Qu 4 . What effect will sport or exercising have on your metabolic condition?
Answer

Qu. 5. Are you going to tell new friends only if you need to, or will you tell them anyway so it maks it easier to be yourself?
Answer

Qu 6. What can you eat at your favourite café?
Answer

Qu 7. What can you eat at takeaway outlets?
Answer

Qu 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?
Answer

Qu 9. What are some of your ideas for quick meals?
Answer

Qu 10. What are some of your ideas for barbecues?
Answer

Q 11. What are some of your ideas for parties?
Answer

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

Qu 13. How do you plan for camping trips?
Answer

Qu 14.How would you manage if you were going away for the weekend with people who didn't know about your diet?
Answer

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

Q 16. What are you going to tell your girlfriend or boyfriend about your metabolic condition? What is it important for them to know?
Answer

Answers

Qu 1. How long should some one with a metabolic condition stay on the low protein diet?

Answer

In one word . . .**forever.** A low protein diet for a metabolic condition is always for life. It may become a little more flexible as you get older, once you have stopped growing. But for many metabolic conditions there is a risk of serious consequences if you overdo protein intake.

If you want to make changes to your diet or supplement then it is — an absolute **must** to talk to your doctor and dietitian. It is important that you understand the effects of not following your diet strictly. Any changes must be done in the best possible way with the fewest of side effects. Your doctor and dietitian will work with you to fit your diet into your lifestyle.

Qu 2. What makes your blood levels of harmful metabolites rise?

Answer

- 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 in chapter 1.

Qu 3. When can you have more protein in your diet ?

Answer

Blood levels of some amino acids can lower if you are not getting the protein you need from food. Protein needs increase at certain times, such as in the teenage years, building muscle by exercising or in pregnancy

Qu 4. What effect will sport or exercising have on your metabolic condition?

Answer

- Exercise is good for your health and how you feel

- When exercising it is important to eat enough to make up for the energy used up.
- Extra exercise won’t bring your blood levels of harmful metabolites down if you have eaten too much protein.
- Exercise that builds muscle may increase the amount of protein you can tolerate in food over time (see Exercise in Chapter 14).

Qu 5. Are you going to tell new friends only if you need to, or will you tell them anyway so it makes it easier to be yourself?

Answer

Not letting on you have a metabolic condition could make managing it more difficult. 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 a metabolic condition?

Just in case you didn’t know, having a metabolic condition 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, your metabolic condition is just one part of you. You have many qualities and your metabolic condition is not the only one that makes you unique.

Qu 6 .What can you eat at your favourite café?

Answer

Write down your ideas on a separate page then turn to Chapter 13 pages 9 and 10 for some more suggestions.

Qu 7. What can you eat at takeaway outlets?

Answer

Write down your ideas on a separate page then turn to Chapter 13 pages 9 for some more suggestions.

Remember that many takeaway food outlets label their food with the nutrient content or provide this information on their websites so you can work out the protein content. And don't forget that commercial vegetable burgers are **not** low in protein.

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

Answer

Write down your ideas on a separate page then turn to Chapter 13 pages 10 to 12 for some more suggestions.

If you eat regularly at a restaurant or know ahead of time where you are going, you'll often find the restaurant happy to cook low protein food.

Qu 9. What are some ideas for quick meals?

Answer

Write down some ideas on a separate page then see Chapter 10, page 11, Food on the Run.

Q 10. What are some ideas for barbecues?

Answer

Write some down on a separate page then go to Chapter 13 pages 8 to 9 for some more ideas.

Qu 11. What are your ideas for parties?

Answer

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 to share. Most people like vege chips, low protein dips and vegetable sticks. (See Chapter 13 for more party food ideas.)

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

Answer

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 if available (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. Don't go too long without it though.

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), low protein bread/buns, snack bars and low protein biscuits and crackers.

Qu 13. What are your ideas for camping trips?

Answer

Take enough of your special supplement for the number of days you will be away and a bit to spare. Supplements that are in sachets or ready-to-drink if available 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.

Plan what food you will take. Plenty of low protein food will help you keep your energy levels up —essential if you are busy outdoors. Turn to Chapter 13, pages 7 and 8.

Qu 14. How would you manage if you were going away for the weekend with people who didn’t know about your diet ?

Answer

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

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.

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

Answer If you are over in your protein intake for one day, you can cut back at the next meal of on the next day. Only go over by 10 per cent or one tenth more than your usual protein if you can manage it. This means that if your usual diet is 20g protein then only go over by 2 g . Do not do this regularly or too often!

Qu 16. What are you going to tell your girlfriend or boyfriend about your metabolic condition? What is it important for them to know?

Answer

It is essential that they know about your metabolic condition, especially if you are spending a lot of time with each other. Important things to tell others are:

- what your metabolic condition is
- what you can eat
- about your supplement
- what to do if you become unwell or have an accident.

Offer them your handbook to look at.

ASIEM food lists for protein counting

The Metabolic dietitian will advise you on how to restrict protein in the diet. The protein in some foods, but not all, may need to be counted each day. To count protein you will need to use information from food labels as well as the lists below.

There are several ways that protein can be restricted from food depending on the level of restriction needed. For your child's diet (or your diet if you have the disorder) you need to do one of the following as advised by your dietitian:

- Avoid food containing protein from animal sources, legumes or nuts (high protein food lists) and count protein from some fruit and vegetables and all other foods that contain more than 0.3g protein per serve **OR**
- Avoid all protein from animal sources or legumes (high protein food lists) but not count other foods such as fruit, vegetables, breads, cereals, pasta and other lower protein foods **OR**
- Include small amounts of protein from animal sources, legumes or nuts using the high protein food lists, but do not count other foods such as fruit, vegetables, bread, cereals, pasta and other lower protein foods.

Whatever the levels of protein restriction the aim is not to count every gram of protein eaten. This method of counting is:

- more practical
- gives good blood test results
- encourages intake of fruit, vegetables and other low protein foods.

Some amino acids (e.g. leucine, methionine, phenylalanine) are in lower amounts in fruits and vegetables compared to other foods, even when the amount of protein is the same. This has been taken into account in these lists.

The prescribed number of grams of protein can go further if::

- Meals and snacks are based around a variety of fruits and vegetables.
- You choose packaged goods (e.g. breakfast cereal, stock) with the lowest protein content. This will allow you to include more foods in your protein allowance.
- You include plenty of low protein foods including the special bread, pasta, etc. These are great 'fillers' on a low protein diet.

Using the protein lists

The food lists provided are for foods that do not come with a label e.g. potato. Use labels where possible on packaged foods to calculate protein content.

It is useful to think of foods as having 3 different levels of protein per serve:

- **Free foods**—these foods contain no or small quantities of protein and if used in normal quantities are usually allowed without restriction. These foods are important as the basis of meals and to give variety. Many fruits and vegetables are free foods. If eaten in large quantities some free foods may cause blood levels of harmful metabolites to rise and may need to be counted. Your dietitian will advise you.
- **Moderate protein foods**—these contain more than 1g protein per serve. These may need to be counted.
- **High protein foods**—these foods contain protein from animal sources, legumes, nuts or seeds, and contain large amounts of protein per serve. These foods are not usually included in a low protein diet unless tolerance to protein is high. Some people may be able to include small quantities but discuss this with your dietitian.

Weighing versus using household measures

These lists give protein content for average serve sizes as well as the amount of food that contains 1 gram of protein. Weighing is likely to be more accurate than household measures.

Standard metric measures are used:

- 1 teaspoon = 5 ml
- 1 cup = 250 ml
- All measures are level

Note that:
in Australia 1 tablespoon = 20 ml
in New Zealand 1 tablespoon = 15ml

How to use the tables

The tables listing protein content of foods on the following tables give information on foods that may not have a label and contain moderate to high amounts of protein per serve. Use food labels when ever possible to calculate the amount of protein in food.

Your dietitian will advise which foods need to be counted.

When counting protein in foods or from food labels always round to the nearest ½ gram of protein:

- Do not count in ¼ grams of protein. Counting in ½ grams is accurate enough. Values in tables have been rounded to ½ gram.
- Round any values you have calculated to the nearest ½ gram:
    - 2.3g protein is counted as 2.5g
    - 1.2g protein is counted as 1g
- A food containing 0.3 g protein or less per serve does not need to be counted. It is a free food.
- Food containing 0.4 g protein or more per serve needs to be counted.
- Check that the serve size given on pack is appropriate—serve sizes vary.

Using the tables
The un-shaded columns in the table give information on the protein content for a typical serve of the food. The serve is described in the first un-shaded column and the weight of that serve is given in the next un-shaded column. The third un-shaded column gives the number of grams of protein in that serve.
The last column of the table gives the weight of the food that contains 1 gram of protein. This value is useful when calculating protein content of recipes or if weighing foods. Foods with the smallest number are highest in protein.

Foods with low to moderate amounts of protein

Free foods contain small quantities of protein and if used in normal quantities are allowed without restriction. These foods are important 'fillers' and to give variety.

Fruit

Healthy eating guidelines recommend at least two (2) serves of fruit a day. Most fresh, frozen, dried or canned fruit contain only small amounts of protein.

Some fruit needs to be counted as these contain more protein than other fruits – see table. Even if you are advised to count these particular fruits you do not need to count them if only a small amount is eaten. For example do not count if less than the following amounts are eaten:

- banana—¼ small
- mulberries—¼ cup
- passion fruit—½ fruit
- pomegranate—1or 2 teaspoons
- strawberries—2 medium or 1 large
- dried apricots or figs—1 or 2 pieces
- dried currants or sultanas—1 or 2 teaspoons.

If you purchase foods in which a free fruit is mixed with another ingredient—such as custard, cereal or breadcrumbs—use the value for protein on pack. For example:

- Canned apple is free but baby canned apple with cereal OR baby

canned apple with custard needs to be counted using the protein value on pack
- Fruit slice biscuits need to be counted using the protein value on pack.

	Type	Serve description	Edible weight of serve	Protein per serve	Weight containing 1g protein
Banana	Common	½	70g (110g with skin)	1g	60g
Banana	Sugar	1	60	1g	67g
Mulberries	Fresh	½ cup	65	1.5g	45g
Passion fruit	Fresh	2	45g of pulp	1.5g	33g
Pomegranate	Fresh	½ fruit	120g (210g with skin)	2.5g	53g
Strawberries	Fresh	5 medium	60g	1g	59g
Tamarillo	Fresh	1	75g	1.5g	50g
Apricot	Dried	5 halves	25g	1g	22g
Fig	Dried	3 figs	45g	1.5g	28g
Sultanas or currants	Dried	Small packet	40g	1g	36g

Vegetables

Healthy eating guidelines recommend five (5) serves of a variety of vegetables each day. Many vegetables do not need to be counted on a low protein diet if eaten in usual serves. Carrots, celery, cucumber, eggplant, green zucchini, lettuce and tomato are particularly low in protein.

Some vegetables may need to be counted as these contain more protein than other vegetables—see table. Your dietitian will tell you which of the following to count:

- potato, peas, corn
- broad beans, green beans
- mushroom, kohlrabi

- broccoli, spinach, Brussels sprouts, cauliflower, bok choy, mustard greens, red cabbage
- asparagus, avocado, globe artichoke, bean sprouts, button squash, mushrooms
- pumpkin, sweet potato

Even if you are advised to count these particular vegetables, you do not need to count them if only a small amount is eaten (e.g. in a stir fry or a ‘taste’).

Dried beans and lentils, including baked beans, dried beans, lentils and chick peas, are too high in protein for most people needing a low protein diet and should be avoided. See high protein food lists on pages 17 to 22.

If you purchase foods in which a free vegetable is mixed with another ingredient such as counted vegetable, rice, pasta or breadcrumbs, use the value for protein on pack. For example:

- Canned carrots are free, but canned carrots with rice or couscous needs to be counted using the protein value on pack.
- Frozen carrots are free but a frozen mix of corn, carrots and peas should be counted using the protein value on pack if you count corn and peas.
- Sun-dried tomatoes are free but a sun-dried tomato pesto with pine nuts and parmesan is counted using the protein value on pack.

	Type	Serve description	Edible weight of serve	Protein per serve	Weight containing 1g protein
Asparagus	Raw, boiled, canned	3 spears	42g	1g	40g
Artichoke, globe	Boiled	½ globe	67g	2g	30g
Avocado	Raw	½	75g	1.5g	50g
Bok Choy	Raw	1 cup	60g	1.5g	38g
Bean sprouts	Raw	½ cup	50g	1.5g	37g
Broad beans	Raw, boiled	¼ cup beans	40g	3g	14g
Beans, green	Raw, cooked	½ cup	60g	1.5g	41g
Broccoli	Raw, boiled	1 cluster	45g	2g	22g
Brussels sprouts	Raw, boiled	4	75g	3g	26g
Button squash	Raw	1 squash	40g	1g	37g
Cabbage	Mustard , red	1 cup shredded	60g	1.5g	43g
Cauli-flower	Raw, boiled	1 cluster	50g	1g	48g
Corn					
Baby corn	Canned	8 spears	130g	2g	62g
Creamed corn	Canned	½ cup	130g	2.5g	50g
Corn kernels	Canned, frozen, boiled	½ cup	85g	2.5g	33g
Corn on the cob	Raw, frozen boiled	½ cob or 1 cobette	75g	3g	24g
Kohlrabi	Raw, boiled	½ cup sliced	75g	3g	26g

	Type	**Serve description**	**Edible weight of serve**	**Protein per serve**	**Weight containing 1g protein**
Mushrooms	Raw	4 small	45g	1.5g	30g
Peas					
Peas, green	Fresh, canned or frozen, boiled,	¼ cup	40g	2.5g	17g
Peas, split	Boiled	¼ cup	40g	3g	15g
Snow peas	Raw, boiled	10 pods	35g	1g	35g
Potato					
Potato	Peeled, raw or boiled or baked in jacket	1 medium or 2 small	120g	3g	40g
Potato	Roasted in oil	1 medium	100g	3.5g	28g
Potato chips	Take-away, oven fried	small serve	70g	3.5g	21g
		6 chips	30g	1.5g	21g
Hash-brown	Fried take-away	2 cakes	110g	2.5g	48g
Potato salad	Bought	1 cup	180g	3.5g	53
Potato scallop	Fried take-away	1 scallop	95g	5g	19g
Pumpkin	Peeled raw, boiled, baked	½ cup	70g	2g	40g
Spinach, English	Frozen or boiled	¼ cup	45g	1.5g	29g
Spinach	Raw	1 cup	35g	1g	38g

	Type	Serve description	Edible weight of serve	Protein per serve	Weight containing 1g protein
English					
Sweet potato, orange	Peeled, raw, boiled or baked	½ cup	120g	2.5g	45g
Taro	Peeled, sliced and boiled	½ cup	60g	1g	50g

If large quantities of free foods are eaten this may affect blood test results. Discuss the range of foods eaten with your dietitian.

Flours, grains, pasta, breads, cereals and biscuits

Most flour, grains, pasta and foods made from them are quite high in protein and need to be counted. Low protein versions are better choices unless protein tolerance is high. Remember that if your child gets used to the taste of ordinary bread, they may not eat the low protein types.

The following can be used freely:

- special low protein flour, baking mixes, breads, biscuits, pasta, rice and breakfast cereals (see Chapter 7)
- arrowroot
- corn flour
- custard powder
- tapioca, sago
- baking powder, baking soda, cream of tartar
- yeast if used to make bread with low protein flour
- food colours and essences e.g. vanilla, almond
- egg replacer (see Chapter 7)
- bean thread vermicelli (check food labels)

Most flours, grains, pasta, bread, cakes and pastry have nutrition labels on the packaging. Use the label where possible to count protein. The following values are a guide only.

Prepared foods—bread, pasta, cakes, biscuits

You may need to count the following:

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Bread and Rolls					
Bread	Fruit loaf	1 slice	33g	2.5g	12g
	Lavash	1	55g	4.5g	12g
	Lebanese	½ bread	55g	5g	11g
	Naan	½ naan	50g	4g	12g
	Tortilla	1	43g	6g	7g
	White	1 slice	30g	2.5g	13g
	White	1 slice, no crusts	20g	1.5g	13g
	Whole meal, brown	1 slice	30g	3g	10g
Bread crumbs	Commercial	½ cup	50g	6g	8g
Crumpet	White	1	50g	2.5g	20g
Muffin	English	½ toasted	40g	4g	10g
Pikelets	Commercial	1	25g	1.5g	17g
Rolls	Hot dog roll	1	100g	10g	10g
	Standard roll	1	65g	6.5g	10g
	Small dinner roll	1	35g	4g	9g
Cakes					
Doughnut	iced	1	80g	4.5g	18g
Lamington	Commercial	1	75g	3.5g	21g
Meringue	Commercial	1 small	20g	0.5g	38g
Noodles					
Rice stick	boiled	½ cup	70g	1g	63g
Wheat Asian style	boiled	½ cup	70g	4.5g	16g
Pasta					
Gnocchi, potato	boiled	½ cup	90g	3.5g	24g
White	raw	½ cup or	26g	2.5g	10g

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
pasta		30 strands			
	boiled	½ cup or 30 strands	70g	3g	24g
Spaghetti	canned	1 small tin	110g	2.5g	43g
Pastry					
Filo	Raw	2 sheets	27g	2.5g	10g
Puff pastry	Raw	½ sheet	85g	4.5g	18g
Shortcrust	Raw	½ sheet	90g	5.5g	19g
Vol-u-vents	Commercial, cooked	1 medium	13g	1g	12g
Snacks					
Popcorn	Commercial	1 cup	15g	1.5g	11g
	Air popped	1 cup	9g	1g	8g
Prawn crackers	Take-away	10 pieces	24g	1g	27g

Ingredients—flours, grains

You may need to count the following:

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Barley, pearl	Boiled	½ cup	95g	2.5g	42g
Bulgar	Boiled	½ cup	133g	3.5g	38g
Bulgar	Dry	¼ cup	45g	4.5g	10g
Bulgar	Soaked	½ cup	102g	5.5g	19g
Coconut	Desiccated	½ cup	40g	2.5g	15g
Couscous	Cooked	½ cup	70g	3.5g	19g
Couscous	Raw	¼ cup	46g	5 g	9g
Flour, white or whole meal	Plain or self raising	½ cup	70g	7.5g	9g
Oat bran	Raw	4 teaspoons	11g	2g	6g

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Polenta	Raw	¼ cup	38g	3g	12g
Potato flour	Raw	½ cup	75g	5g	15g
Rice	White, raw	¼ cup	51g	3g	16g
	White, boiled	½ cup	95g	2g	50g
	Brown, raw	¼ cup	49g	3.5g	14g
	Brown, boiled	½ cup	90g	2.5g	34g
Rice flour	Raw	½ cup	83g	5g	18g
Rolled oats	Raw	¼ cup	24g	2.5g	9g
Rye whole flour	Raw	½ cup	55g	7g	8g
Semolina	Dry	¼ cup	40g	4g	10g
Wheat bran	Raw	8 teaspoons	10g	1.5g	6g
Wheat germ	Raw	4 teaspoons	6	1.5g	4.5g

Seasonings, stocks, sauces and condiments

The following are low in protein and do not usually need to be counted in normal amounts:

- salt, pepper, herbs and spices
- curry powder and pastes (check food labels)
- salad dressing e.g. French, Italian, coleslaw dressing, 1000 island dressing, mayonnaise (check food labels)
- tomato, barbecue and Worcestershire sauce
- fruit chutney, mustard pickles
- mustard
- gravy powder and ready made gravy (check food labels)—**do not**

make gravy with pan drippings

- ready made pasta and casserole sauces (check food labels)
- stock cubes and stock (check food labels for lowest). **Do not make stock from bones or meat.**
- vinegar

You may need to count protein from the following foods:

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Coconut	Fresh	1 piece 3x3x2 cm	30g	1g	30g
Low protein cheese	Loprofin™ cheddar and mozzarella		30g	1g	28g
Marmite™, Bonox™		1 teaspoon	6g	1g	6g
Meat paste	Commercial	1 teaspoon	5g	1g	8g
Peanut butter		1 teaspoon	6g	1.5g	4g
Soy sauce		4 teaspoon	24g	1.5g	19g
Vegemite™		1 teaspoon	6g	1.5g	4g

Fats and Oils

Most fats and oils are very low in protein but cream contains moderate amounts. Some reduced fat versions of butter and margarine may also be higher in protein so check the label if you choose these.

The following contain very little protein and may be used freely:

- butter and margarine
- copha
- oil, cooking fat, ghee
- aerosol and whipped cream with less than 0.3g protein per serve.

You may need to count the following:

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein

Coconut cream	Canned	¼ cup	62g	1g	62g
Cream	Light sour	4 teaspoons	21g	1g	26g
	Pure, UHT or thickened	8 teaspoons	40g	1g	43g
	Reduced fat	6 teaspoons	30g	1g	33g
	Rich or double	12 teaspoons	60g	1g	63g
	Sour	8 teaspoons	40g	1g	43g
	Whipped	12 teaspoons	30g	1g	29g

Drinks

If your child is on a **supplement** it is important that the prescribed amount is drunk each day. Other drinks can include water (which is the best thirst quencher), substitutes for milk and limited amounts of juice, soft drinks and cordials. During illness fluid needs change (see Chapter 3).

Milk is high in protein so **low protein milk substitutes** are useful for cereal, to make low protein drinks, custards and puddings. If these substitutes contain more than 0.3g protein per serve you may need to count them. Suitable low protein substitutes for milk include:

- Rice or oat drinks from supermarkets (long life milk section) and health food shops. Check label for the lowest protein content.
- A mixture of cream and water (1 tablespoon + 100ml water)
- Vance’s DariFree™, Duocal™, Loprofin PKU low protein tetrapak™, Milupa lp drink™. Ask your dietitian how you can get these.

Water is the best thirst quencher. Too much juice, soft drink, and cordial can dull a child’s appetite for food and are not good for teeth. These drinks are best kept to being a treat once or twice a week or for when ill. Juice should be diluted for babies and limited to 100ml per day. Most drinks are low in protein:

- Cordials, soft drinks, mineral and soda water (plain and flavoured) contain no protein
- Some juices contain more protein that others. Limit 100% juice to one cup a day for older children. It is better to drink water and eat the fruit!
- Tea (black, green, herbal) and coffee without milk does not need to be counted for older children and adults
- Flavoured 'milk' drinks can be made using low protein milk substitutes and low protein flavourings such as Nesquik ™ (not Nesquik Plus™) or Sippah flavoured straws™ (check protein content)

These drinks may need to be counted:

	Type	**Serve description**	**Weight of serve**	**Protein per serve**	**Weight containing 1g protein**
Cocoa	Powder	4 teaspoons	7g	1.5g	5g
Milk	Full cream	½ cup	125ml	4g	30ml
Milo™, Ovaltine™, Malted milk	Powder	4 teaspoons	7g	1g	8g
Nesquik Plus™	Powder	5 teaspoons	10g	1g	12g

Alcoholic drinks are NOT suitable for children but can be used in cooking such as casseroles.

Beer, stout and milk or cream based liqueurs need to be counted (see table). The following drinks have little or no protein:

- wine—red, white, champagne, wine coolers
- port, sherry
- spirits—such as rum, whiskey, gin, vodka.

Check Chapter 14 for guidelines on responsible drinking for adults. Remember that these guidelines refer to standard drink sizes. Alcoholic drinks are often purchased in larger sizes than one standard drink so some drinks can add up to a significant amount of protein as well as alcohol:

	Serve	Serve size	Protein per serve
Beer, extra-lite beer, lite beer	1 can	375 ml	1g
Stout	1 bottle	375 ml	2.5g
Advocaat or cream based liqueur		30ml	1g

Sugar, desserts and lollies

Sugar is naturally low in protein but other ingredients can increase the protein content of lollies and desserts. For example jelly beans and jelly snakes can vary from 0 to 6 g protein per 100g.

The following contain very little protein and may be used freely:

- sugar (all types)
- jam, honey, golden syrup, maple syrup, marmalade, treacle
- jelly thickened with vegetable gum (not gelatine). Check the ingredient list: the numbers 406, 407, 410, 412, 413, 414, 415, 416 are vegetable gums. Ready made jellies (in the refrigerated dessert section of the supermarket) are usually set with vegetable gum.
- lollies without chocolate, milk, gelatine or nuts—check the protein content of barley sugar, boiled lollies, toffee, jelly beans, pastilles, snakes, candy floss, fairy floss, plain honeycomb. Take care if you have tyrosinaemia to check for aspartame in sugar free lollies.
- low protein chocolate (Duobar™, Vita-bite™, Loprofin crunch bars™)—ask your Dietitian where you can buy these.
- ice blocks without ice cream or milk e.g. icy poles
- ice cream toppings (check protein content), 100’s and 1000’s sprinkles.

Frequent eating of high sugar foods and drinks can cause tooth decay. See Chapter 10 for advice on dental care.

The following table shows protein values of some higher protein lollies and desserts. Low protein alternatives are recommended (such as jellies set with vegetable gum, lower protein gelato and sorbets).

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Chocolate	Cooking	6 squares	29g	1g	25g
	Dark	6 squares	29g	1.5g	20g
	Milk, white	6 squares	29g	2.5g	12g
Gelatine	Regular	4 teaspoons	13g	11g	1g
Ice cream	Vanilla, regular	2 scoops	100g	4g	25g
Jelly crystals	Gelatine based	1 packet	85g	6g	14g
Jelly made up	Gelatine based	1 cup	280g	3g	100g
Licorice		10cm piece	26g	1.5g	20g
Marshmallow	No chocolate	6	27g	1g	26g
Marzipan	No chocolate	1 cm slice	15g	1g	14g

High Protein foods

Your dietitian will advise whether any of these foods can be included in the low protein diet.

Beans and legumes

Most dried beans and legumes contain about 25g protein per 100g. With cooking or canning the water content increase so there is less protein per 100g.

Beans and legumes naturally have a lower methionine content compared to some other foods. Discuss this with your dietitian. It may be possible to include these foods in a low protein diet for homocystinuria.

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Baked Beans	Canned	½ cup	138g	6.5g	22g
Butter beans	Fresh, boiled	½ cup	80g	2g	40g
Cannellini beans	Canned, drained	¼ cup	64g	4g	16g
Haricot beans	Dry	¼ cup	50g	10g	5g
	Boiled	¼ cup	44g	4g	12g
Lima beans	Dry	¼ cup	56g	11g	5g
	Boiled	¼ cup	50g	3g	16g
Mixed beans	Canned, drained	¼ cup	50g	3g	16g
Red kidney beans	Dry	¼ cup	45g	10g	4.5g
	Canned, drained	¼ cup	48g	3g	15g
	Boiled	¼ cup	45g	3.5g	12.5g
Soya beans	Canned,	¼ cup	49g	4g	12g

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Soya beans	Boiled	¼ cup	43g	6g	7.5g
Soya beans	Dry	¼ cup	46g	15g	3g
Chickpeas	Canned, drained	¼ cup	46g	3g	16g
	Hommus	4 teaspoons	30g	3g	11g
Lentils	Boiled	¼ cup	48g	3g	15g
	Dry	¼ cup	48g	12g	4g

Nuts and seeds

Most nuts and seeds contain about 20g protein per 100g. However some such as chestnuts, macadamia nuts and pecan nuts are lower in protein and may be useful to include in pesto, toppings or baking.

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Almonds	Blanched or with skin	20 nuts	20g	4g	5g
Brazil nut	Raw or blanched	5 nuts	20g	3g	7g
Cashew	Raw or toasted	8-12 nuts	20g	3.5g	6g
Chestnuts	Raw	2 small	20g	0.5g	41g
Hazelnuts			20g	3g	7g
Macadamia nuts		5 medium	20g	1.5g	13g
Peanuts	Raw, toasted	25 nuts	20g	5g	4g
Peanut butter	Paste	4 teaspoon	25g	6g	4g
Pecan nuts	Raw	9 halves	20g	2g	10g
Pine nuts	Raw	4 teaspoon	14g	2g	8g

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Sesame seeds		4 teaspoon	13g	3g	4.5g
Sunflower seeds		4 teaspoon	16g	3.5g	4.5g
Tahini	Paste	4 teaspoon	21g	4g	5g
Walnuts	Raw	12 halves	20g	3g	7g

Meat, chicken and fish

Raw lean beef, chicken, fish, kangaroo, kidney, lamb, liver, pork, rabbit, turkey and veal all contain about 20g protein per 100g i.e. 5g raw meat contains 1g protein.

With cooking meat dries out so that the protein content per 100g increases. Depending on the cooking method 3-4g cooked meat contains 1g protein.

Even fat and skin contains 10-15g protein per 100g so using fattier cuts of meat still provides significant amounts of protein.

Meat, chicken or fish serves will look bigger if you:

- slice it thinly
- stir-fry or casserole it with lots of vegetables
- make kebabs with vegetables for the BBQ.

The table on the next page gives protein content in average serves of meat. Remember that one person’s idea of a serve of steak or chicken or a slice of ham can be very different from another person’s. It is worth working out the size of the usual meat, chicken or fish serving your child eats from the amount you buy. If you buy packaged meat such as ham, luncheon meat, salami, chicken nuggets, fish fingers, work out the amount of protein in one slice or piece from the pack.

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Beef					
Mince	Raw	¼ cup	60g	12g	5g
Topside	Raw	1 slice	50g	11g	5g
Deli meats					
Bacon	Grilled	1 breakfast rasher	35g	8g	4.5g
Bacon fat	Grilled, no rind		30g	4g	7g
Frankfurt		1 cocktail size	25g	3g	8g
		1 hot dog size	75g	9g	8g
Ham	From leg	1 thin slice	10g	2g	5.5g
Luncheon meat	e.g. Devon	1 slice	25g	3g	8.5g
Salami		2 thin slices	10g	2g	4.5g
Sausage	Raw	1 thin or ½ thick	55g	7g	8g
Chicken					
Breast	Raw	¼ breast	56g	12.5g	4.5g
Drumstick	Baked	1	30g	8g	4g
Skin	Baked		30g	7g	4.5g
Thigh	Baked	1 (skin and meat)	100g	24g	4g
Wing	Baked	1	30g	9g	3g
Nuggets	Cooked	6	125g	12g	10g
Fish and Seafood					
Anchovies	Canned	1 anchovy	4g	1g	4g
Calamari/ squid	Raw		100g	17g	6g
Crabmeat	Canned	Small tin	100g	12.5g	8g
Fish	White, raw	½ bream fillet	87g	17g	5g
Fish finger		3	75g	7.5g	10g

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Fish battered		1 piece	150g	28g	19g
Mussel, green	Flesh, cooked	6	48g	9g	19g
Prawn, king	Raw	2	40g	8g	5g
	Cooked	2	35g	8g	4g
Salmon or tuna	Canned	Small tin	100g	20g	5g
Salmon or trout	Smoked, sliced	1 slice	25g	6g	4g
Tuna sashimi		1 serve	95g	25g	4g
Lamb					
Chump chop	Grilled	1	60g	20g	3g
Loin chop	Grilled	1	50g	14g	3.5g
Cutlet		1	35g	10g	3.5g
Leg	Roast	1 slice	30g	9g	3.5g
Pork					
Crackling	Roast	½ cup	50g	17g	3g
Leg	Roast	1 slice	45g	11g	4g
Loin chop	Grilled	1	200g	54g	4g
Schnitzel	Pan fried	1	200g	57g	3.5g
Veal					
Cutlet	Grilled	1	140g	39g	3.5g

Milk, cheese, eggs and dairy food

Use lower protein alternatives for milk and soy milks (see page 14). Ice blocks, gelato, sorbet and fruit based ice confections are usually a lower protein choice than standard ice cream or yogurt.

Block cheese contains 25-30g protein per 100g. Low protein cheese is available (see page 13). Cream cheese and cheese spreads may be able to be included on a reduced protein diet (check protein content on pack).

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Cheese					
Cream cheese		4 teaspoon	20g	1.5g	12.5g
Cheddar		1 slice	21g	5.5g	4g
Ricotta		4 teaspoon	20g	2g	10g
Egg					
Whole	Raw	1 x 60g egg	50g	7g	7.5g
White	Raw	1 white from 60g egg	30g	3.5g	9g
Yolk	Raw	1 yolk from 60g egg	20g	3g	6.5g
Ice cream	Vanilla	2 scoops	100g	4g	25g
Milk	Full cream	½ cup	125ml	4g	30ml
Soy drink		½ cup	125ml	3.5g	38ml
Yogurt	Fruit	1 tub	200g	9g	22g
	Natural	1 tub	200g	11g	18g

Eating away from home

Some of the choices in the tables below will use up a lot of the daily protein allowance. See Chapter 13 for alternatives. Many of the fast food chains have nutritional information available in store or on their websites— check these for suitable choices.

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Fast food					
Chicken burger or hamburger	With mayonnaise and lettuce	1	190g	24g	8g
Chiko roll	Deep fried	1	170g	13g	13g
Calamari,	Crumbed, fried	3 rings	66g	12g	5.5g
Fish	Battered and fried	1 piece	150g	21g	7g
Fish Cake	Deep fried	1 cake	80g	9g	8.5g
Fish cocktail	Deep fried	1	30g	4.5g	7g
Fish or seafood stick		1	27g	3g	9g
Meat pie	Individual size	1	190g	15g	13g
	Party pie	1	40g	3.5g	12g
Onion rings	Battered and fried	6 medium rings	36g	1.5g	23g
Pasty	Meat and potato	1	165g	11g	15g
Pizza	Ham and pineapple or supreme	¼ medium size	130g	16g	8g
Potato chips		Small serve	90g	4.5g	21g
Potato scallop	Fried	1 scallop	95g	5g	19g

	Type	**Serve description**	**Weight of serve**	**Protein per serve**	**Weight containing 1g protein**
Potato salad		1 cup	180g	3.5g	53g
Sausage roll	Individual size	1	130g	10.5g	12.5g
	Party size	1	40g	3.5g	11g
Chinese, Thai, Vietnamese					
Chow Mein (with noodles)	Chicken, beef or pork	1 serve	400g	31g	13g
Dim sim		1	50g	5g	10g
Fish ball		1 small	12g	1.5g	8g
Fried rice noodle	Indonesian	1 serve	400g	29g	14g
Prawn crackers		10 pieces	24g	1g	27g
Rice	White, boiled	1 cup	190g	4g	50g
	Fried	1 cup	165g	10.5g	16g
Spring Roll		1 small	25g	2g	12g
Sweet and sour	Pork or prawns	1 serve	400g	31g	13g
Mediterranean					
Cabbage roll	With meat and rice	1	85g	4g	22g
Kibbi	Vegetarian	1 piece	80g	5.5g	15g
Falafel		2 patties	50g	4.5g	11g
Ravioli or lasagne	With meat	1 cup	250g	20g	12.5g
Spanakopita		1 piece	110g	10g	11g
Tabouleh		¾ cup	100g	2g	42g
Vine leaf stuffed	With meat and rice	1	35g	1.5g	25g
Zucchini stuffed	With meat and rice	1	200g	12.5g	16g
Indian					
Aloo Muttar	Potato & peas	1 serve	400g	28g	14.5g
Dahl		1 serve	60g	10g	6.5g

	Type	Serve description	Weight of serve	Protein per serve	Weight containing 1g protein
Pakora		1 serve	205g	37g	5.5g
Pappadum		1 large	50g	5.5g	9g
Samosa	Vegetable	1	145g	11g	13g

The protein values in these exchange lists are from Australian food composition data (NUTTAB 2006). You can check protein values per 100g of individual Australian foods on line at http://www.foodstandards.gov.au/monitoringandsurveillance/nuttab2006/onlineversionintroduction/index.cfm.

Discuss any queries with your dietitian.

Glossary

Amino acid

Amino acids are the basic building blocks of proteins. The body makes many amino acids. There are also essential amino acids which must be obtained from food.

Anabolism

The building up of food into protein for muscles. Anabolism helps keep metabolic conditions under control.

Branched chain amino acids

Leucine, isoleucine, valine are branched chain amino acids (so called because of their chemical structure). These are not able to be broken down in the usual way in Maple Syrup Urine Disease (MSUD).

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.

Carnitine

An amino acid-like substance produced in the body and found in food. It is important in energy metabolism and can lower levels of harmful metabolites in some inborn errors of metabolism.

Catabolism

The body breaks down its own protein for example in muscles, this often happens during an illness, fever or rapid weight loss. It causes blood levels of harmful metabolites to increase in metabolic conditions.

Cobalamin

This is another name for Vitamin B_{12} which has many functions in the body. It works as a cofactor for some enzymes and may be used in the treatment of methyl malonic aciduria (MMA).

Cofactor

A chemical compound (often a vitamin) which acts as a helper for an enzyme thus improving its function.

Coma

Loss of consciousness. Some of the inborn errors of metabolism can result in coma if control of the condition is lost during illness.

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.

Fasting

Not eating or drinking for a period of time. In metabolic disorders fasting can increase levels of harmful compounds particularly during illness.

Fat

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

Folic acid

A vitamin also known as folate with many functions in the body. It is recommended for all pregnant women to help prevent birth defects.

Formula

Usually refers to a breast milk substitute used for feeding babies with bottles. Special formulae are used for babies for many metabolic conditions. These may be protein free or contain special protein without the amino acid that cannot be broken down. These formulae also contain vitamins and minerals.

Gastrostomy

A semi-permanent tube through the abdomen into the stomach used to give liquid food if food intake is inadequate. It is less visible and more comfortable than a nasogastric tube (see also) but needs a small operation to insert it.

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.

Glutaric aciduria Type 1 (GA1) (sometimes called glutaryl CoA dehydrogenase deficiency)

A disorder of protein metabolism classed as an organic acid disorder.

Gram

One thousandth of a kilogram. A system of counting grams of protein is used to keep track of protein intake.

Guthrie card

The name sometimes given to the card used for recording blood spots from the finger or heel. The Guthrie test was the first newborn screening test introduced in Australia and New Zealand to diagnose PKU.

Homocystinuria

A disorder in the metabolism of the amino acid methionine. Levels of homocystine increase in the blood and urine.

Hormone

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

Hypotonia

Floppiness due to decreased muscle tone. Babies with hypotonia may feel like a ‘rag doll’ when held.

Hyperornithinaemia

Also known as gyrate atrophy; this is a disorder in the metabolism of ornithine which can lead to blindness.

Inborn error of metabolism

An inherited (genetic) disorder of metabolism—a metabolic disorder.

Isovaleric acidaemia (IVA) (sometimes called isovaleric aciduria)

A protein metabolism disorder classed as an organic acid disorder.

Kilojoule

A kilojoule is a measure of energy. One Calorie = 4.2 kilojoules.

Maple syrup urine disease (MSUD)

A disorder of protein metabolism involving leucine, isoleucine and valine metabolism. Named because of the sweet smell of the urine when blood levels of these amino acids are high (but nothing to do with the food maple syrup).

Metabolism

Chemical reactions that occur in the body which are essential for life. It covers the breakdown of food to provide energy and the breakdown of complex substances to simpler ones for use in the body e.g the breakdown of proteins to amino acids.

Metabolic crisis or decompensation

A loss of the usual chemical balance within cells of the body. This usually has a rapid onset and is a medical emergency requiring urgent treatment. Untreated it can be fatal or leave a child with permanent physical or brain damage.

Metabolic Team

Metabolic 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. The team advises on the care of the metabolic condition in outpatient clinics or on admission to hospital.

Metabolite

A substance made from the breakdown of nutrients and chemicals produced in the body during metabolism. Some metabolites are harmful if in greater concentration than usual.

Methylmalonic acidaemia (MMA) (sometimes called methylmalonic aciduria)

A protein metabolism disorder classed as an organic acid disorder.

Nasogastric feeding

Feeding through a tube passed through the nose into the stomach—this is used if food intake is inadequate.

Newborn screening

All newborn babies in Australia and New Zealand have a blood test within the first few days for metabolic conditions among other things.

NTBC

A medication used in the treatment of tyrosinaemia type 1. The full name is 2-(2-nitro-4-trifluoromethylbenzoyl)-1,-3-cyclohexanedione). It is not useful in the treatment of tyrosinaemia type 2.

Organic acid disorders (organic acidurias or organic acidaemias)

A group of disorders resulting from the body's inability to break down particular amino acids. This includes methylmalonic aciduria (MMA), propionic aciduria (PA), maple syrup urine disease (MSUD), isovaleric aciduria (IVA), glutaric aciduria type 1. These conditions are diagnosed when increased levels of different organic acids are found in the blood and urine.

Phenylketonuria

Also known as PKU, this is an inherited condition where the body lacks the enzyme phenylalanine hydroxylase needed to break down the amino acid phenylalanine in foods. Treatment is a low protein diet.

Plunket Nurse

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

PKU

See phenylketonuria

Propionic acidaemia (PA) (sometimes called propionic aciduria)

A protein metabolism disorder classed as an organic acid disorder.

Protein

Protein is made up of amino acids. It is needed by the body for growth, repair and important components like hormones, antibodies, enzymes and blood clotting. Many foods contain protein. Foods such as meat, chicken, fish, eggs, milk, legumes and nuts are rich in protein and not suitable for people on low protein diets—some may be allowed in very small amounts. A special protein supplement is used in some metabolic conditions to make up for the protein unable to be eaten in food.

Protein supplement

A special type of supplement used in the management of metabolic conditions. It contains all the essential amino acids (except the ones not tolerated in the particular medical condition) and may contain vitamins and minerals.

Pyridoxine

Also called vitamin B_6 this acts as a helper for some enzymes and may be used in high doses in hyperornithinaemia and homocystinuria if people are responsive.

Supplement

There are several types of supplements used by people with metabolic conditions to replace some of the protein in their diet and provide energy, vitamins and minerals.

Tyrosinaemia

An inborn error of tyrosine metabolism. Type 1 and Type 2 have very different symptoms.

Urea cycle disorders

A group of metabolic disorders involving the urea cycle. This cycle is responsible for ridding the body of ammonia produced from nitrogen in the breakdown of protein.