



Metabolic.ie

National Centre for Inherited Metabolic Disorders



Urea Cycle Disorder

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Introduction

The aim of this guide is to provide information for individuals with a Urea Cycle Disorder or for parents/carers with a child who has a Urea Cycle Disorder. We hope you find it useful. The information in this handbook is correct at the time of printing and the recommendations made are based on current knowledge. We recommend you use this handbook only under full dietary and medical supervision.

Please find further information on our website www.metabolic.ie

Chapter 1

What is a Urea Cycle Disorder...?



A Urea Cycle Disorder is an inherited metabolic disorder which affects the breakdown of protein containing foods.

Protein is one of the main nutrients in our diet. It is made up of 20 different building blocks called **amino acids**. When protein is eaten it is broken down by **enzymes** into **amino acids**. These amino acids carry out a variety of functions within the body including growth, repairing cells and tissues as well as making other body proteins.

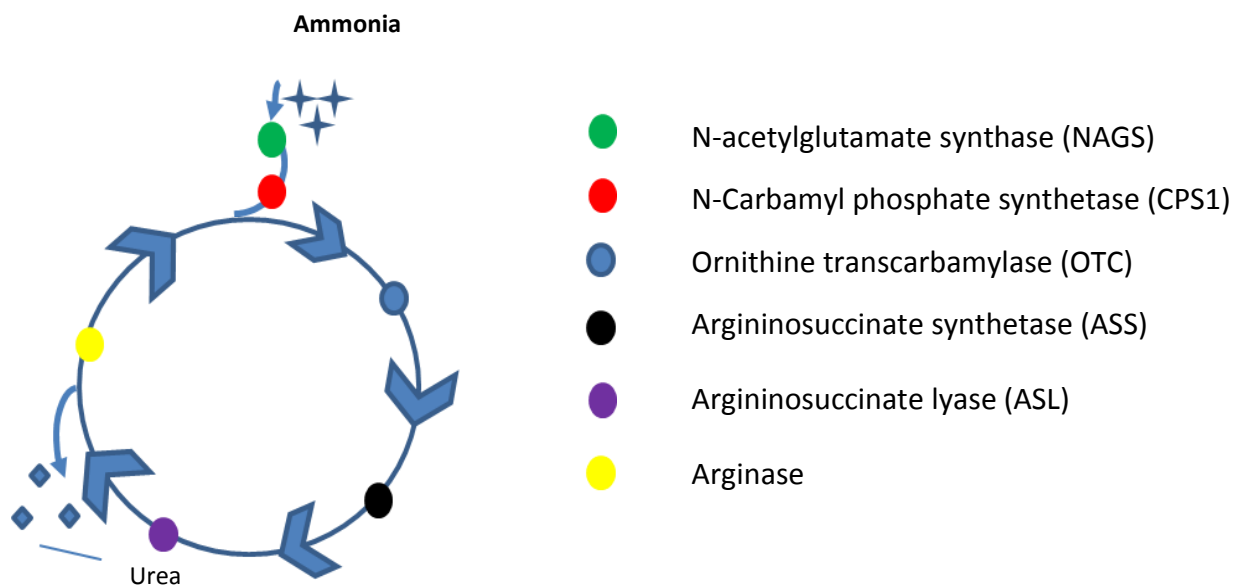
Generally, we consume much more protein than the body needs. Normally, the extra protein that the body does not need is converted in the liver to a chemical called ammonia and then, through a series of steps (known as the urea cycle) into another chemical called urea. Urea will pass through the kidneys where it is removed from the body in the urine. People with a Urea Cycle Disorder are born without or with very little of one of the six enzymes that breakdown ammonia. In Urea Cycle Disorders, the extra protein the body does not use is converted into ammonia, but the liver cannot convert some or all of the ammonia into urea for excretion in the urine. A deficiency of one of the enzymes in the urea cycle results in high levels of ammonia building up in the blood. High levels of ammonia are toxic and it can enter the brain and damage it. If left untreated, high ammonia levels can be life threatening.

In a person **without** a Urea Cycle Disorder, ammonia is converted into urea.

In a person **with** a Urea Cycle Disorder, the conversion of ammonia into urea does not happen as it should and blood ammonia rises which is toxic.

The name of your child's Urea Cycle Disorder is _____.

The name tells you which enzyme isn't working in your child's urea cycle. The following diagram shows the different enzymes involved in the conversion of ammonia to urea.



How is a Urea Cycle Disorder treated?

The main aim of treatment is to ensure acceptable levels of ammonia and amino acids and to provide adequate protein to support normal growth and development. **The treatment of Urea Cycle Disorder is a low protein diet for life** which allows just enough protein for growth and repair of body tissues. Medications to help eliminate ammonia will also be advised by the medical team. The medications are used in different combinations and are specific to your child's needs and their type of Urea Cycle Disorder. In addition, essential amino acids (synthetic protein), vitamins and minerals may be recommended.

An essential part of the treatment of a Urea Cycle Disorder is to take blood samples regularly to measure ammonia and amino acid levels in the blood. These bloods are sent to the Metabolic Lab in Temple Street.

When your child is sick or has high ammonia levels, diet management is different and you will need to ring the metabolic team for advice (see chapter 7 on Urea Cycle Disorder and illness). Medications may also need to change. Your child may need to go to hospital during these times.

How is a Urea Cycle Disorder diagnosed and what causes it?

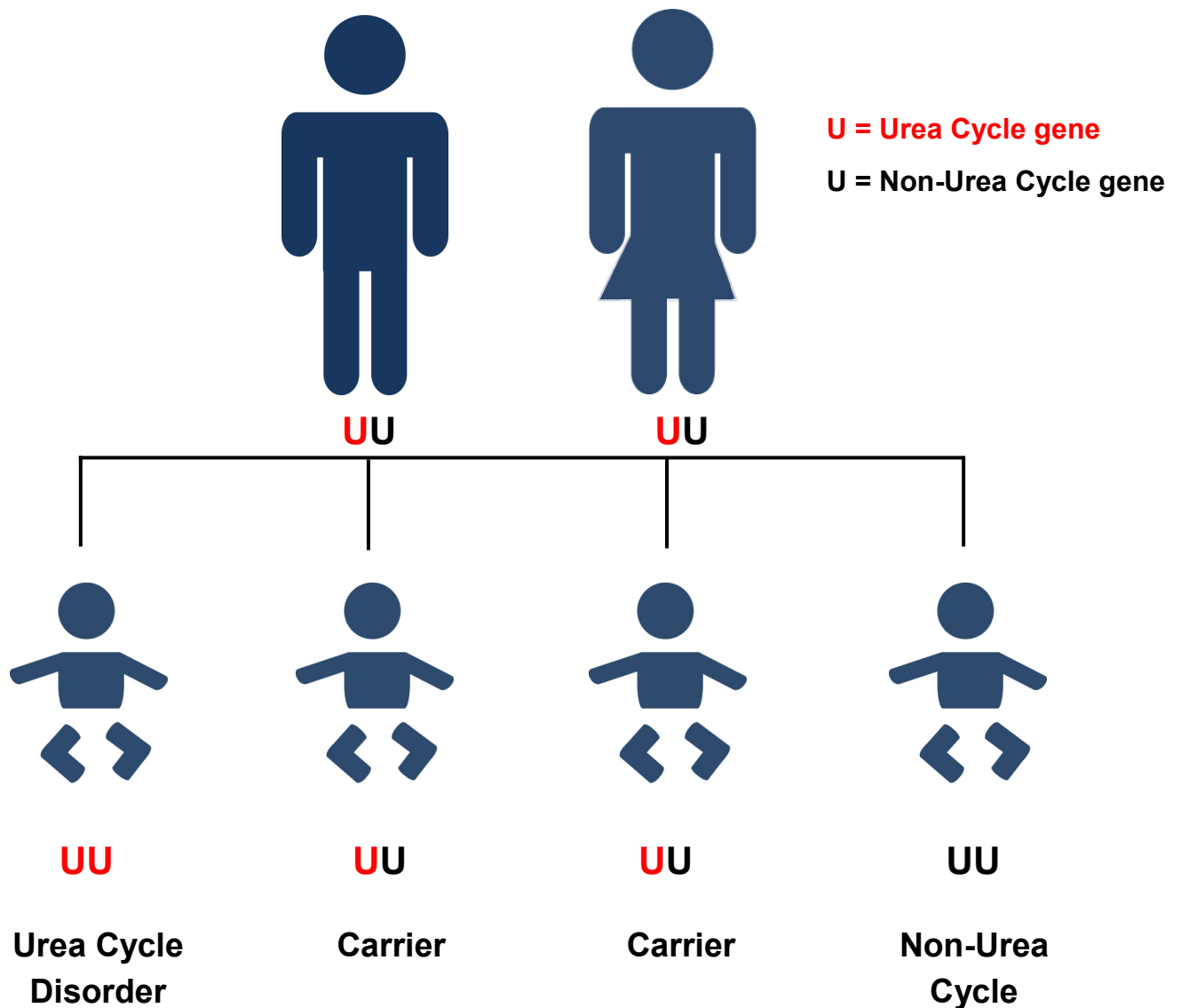
Around one in 8,500 children born worldwide have a Urea Cycle Disorder. The onset and severity of Urea Cycle Disorders is highly variable. This depends on the specific enzyme involved and also on the ability of that enzyme to work. Severe forms result in none or very little enzyme activity and result in severe Urea Cycle Disorders. Mild to moderate forms result when there is some enzyme activity.

Urea Cycle Disorders are inherited metabolic disorders.

Urea Cycle Disorders are diagnosed in both adults and children. Newborn babies with severe Urea Cycle Disorders become severely ill shortly after birth. Children with less severe Urea Cycle Disorders may be diagnosed during childhood or can remain undiagnosed because symptoms are not recognised. Adults often go undiagnosed because they have mild Urea Cycle Disorders which allow them to remove enough ammonia until a stress (e.g. illness) interferes with the ability of the enzyme to work or causes large amounts of ammonia to be made. These adults may have had very subtle symptoms in their lifetime that go unrecognised or unheeded.

1) For Urea Cycle Disorders except for OTC deficiency

Children inherit two sets of genes, one from each parent. As a parent of a child with Urea Cycle Disorder, you have one Urea Cycle Disorder gene and one non-Urea Cycle Disorder gene. This is known as being a carrier. People who are carriers for Urea Cycle Disorder do not have Urea Cycle Disorder themselves. Your child has inherited two Urea Cycle Disorder genes, one from their mother and one from their father. This means that your child has a Urea Cycle Disorder. **It is important to remember a Urea Cycle Disorder is not something you could have avoided and there is nothing you could have done to prevent this.** There is a 1 in 4 chance that another child born to the same parents will have a Urea Cycle Disorder.



2) For OTC deficiency

A person's gender or sex is determined by the so-called sex chromosomes. They exist in two types, X-chromosomes and Y-chromosomes. A female has two X-chromosomes and a male has one X and one Y. One of the more common urea cycle disorders is called OTC deficiency, this stands for ornithine transcarbamylase deficiency. It is inherited as an X-linked disease. This means that this type of disorder is passed on from parents to baby through the X chromosome. If the baby is a female, because females have two X-chromosomes, the good X can balance out the problem with the faulty X. But because males only have one X chromosome, it cannot be balanced by a normal X as in the female and, as a result, in males this disease tends to be more severe.

Occasionally, a child may develop a urea cycle disorder that is not inherited from parents. In such cases, the risk of any future siblings developing the disorder is extremely small.

Chapter 2

Overview of Treatment



The National Centre for Inherited Metabolic Disorders (NCIMD) is where all children diagnosed with Urea Cycle Disorders are treated in the Republic of Ireland. At present, Urea Cycle Disorders are treated by diet and medications.

Successful treatment of Urea Cycle Disorders involves the following:

Foods to avoid: High protein foods such as meat, fish, chicken, eggs, milk, cheese, yogurts, soya, nuts, bread, pasta and chocolate are generally too high in protein and are not allowed in the diet. However, small amounts of these foods may be included in your child's diet depending on their tolerance. This will be discussed with you by your metabolic doctor and dietitian.

Natural protein/Exchanges: People with Urea Cycle Disorders still require a small amount of protein for growth and development; this is what we call their natural protein (exchanges). An exchange is the amount of an allowed food which contains 1 gram of protein. The amount of exchanges allowed will change depending on your child's blood results. Typical exchange foods that are used include measured amounts of potato, beans and cereals.

Protein free foods: Some foods are naturally low in protein and are allowed freely in your child's diet. Examples include most fruit and vegetables, butter/margarine and jam/honey. There are also a number of specially manufactured low protein foods such as bread, pasta and milk which are available on prescription and can be ordered from your local pharmacy.

Daily intake of synthetic protein: A prescribed amount of synthetic protein which contains essential amino acids *may* be recommended. It can be given in the form of a formula, drink or gel. The amount required will be calculated specifically for your child's needs.

Vitamin and mineral and essential fat supplements: These supplements may be recommended to ensure all your child's nutritional needs are met.

Medications: Medications that help to eliminate ammonia will also be advised by the medical team.

Blood levels: Regular monitoring of your child's blood ammonia and amino acids are an essential part of the overall management of Urea Cycle Disorders. Your metabolic team will let you know how often blood levels need to be done and this will depend on your child's age.

Extra care during illness: The metabolic team will advise you on how your child's diet and medications need to change with illness. Each child with a Urea Cycle Disorder will have an individual unwell plan. You need to carry emergency telephone numbers for the metabolic team with you at all times. Your child can become unwell quite quickly so it is better to be overcautious and ring if you are concerned. If your child needs to go to hospital, it is important to take a supply of products for making up unwell feeds and medications. You should also take a copy of the emergency letter from the metabolic doctor and a copy of the unwell plan from the metabolic dietitian.

Keeping supplies up-to-date: It is very important to keep an up-to-date supply of the low protein foods, vitamin and mineral supplements, medications, synthetic protein (if applicable), products for the unwell plan and bloods forms.

Clinic visits: When your child is first diagnosed you will attend the NCIMD regularly for information and support. During clinic, you will meet different members of the metabolic team, for example the dietitian, nurse, doctor, social worker and psychologist. Visits can vary from 6 weekly to 6 monthly. As your child grows older, the time between clinic visits will increase depending on their needs.



Chapter 3

Feeding Your Baby



The first few days

Babies born with Urea Cycle Disorders will have high ammonia levels initially. To reduce ammonia levels, your baby will be given a **protein-free formula** instead of breastfeeds or standard infant formula for the first few days after diagnosis. This will allow the ammonia level to come down quickly. Babies usually make the change to the new formula quite easily. Most babies quickly learn to cope with the combination of bottle and breastfeeding or the different formulas. These should always be given separately and not mixed together.

If your baby is unwell initially and unable to manage feeds by mouth, your baby may need to be given the protein free formula through a tube (called a nasogastric tube) that passes through the nose and into the stomach.

Breastfeeding

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

You can breastfeed your baby and keep the ammonia at an acceptable level. Breast milk **alone** contains too much protein for babies with a Urea Cycle Disorder. Therefore, protein-free formula needs to be given in addition to breast milk. Initially, you will need to express regularly to keep up your supply of breast milk. This is only for a few days until breast feeds can be reintroduced. You will be advised by the metabolic team on how much protein-free formula to give. Also, some babies will be started on a set amount of **synthetic protein** to provide essential amino acids. This will be discussed in more detail in chapter 4. After the protein-free formula and the bottle of synthetic protein (if given), alternate the breast you offer first. If your baby is not interested in the second breast, do not worry, offer this one first at the next breastfeed. Measured amounts of protein-free formula and the synthetic protein (if given) are given by bottle first to reduce the amount of protein taken from breast milk.

A measured amount of protein-free formula +/- a measured amount of synthetic protein are given first and then breastfeed to appetite.

If your baby is hungry in between feeds you may give extra breastfeeds in addition to the measured amounts of protein-free formula +/- synthetic protein.

While breastfeeding, you do not need to alter your diet in any way because your baby has a Urea Cycle Disorder. However, the following points may help you during this time:

- 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 fizzy drink intake
- Breastfeeding mothers need to drink an extra three to four glasses of fluid a day – water is best for quenching thirst
- Your milk supply will adjust to your baby's feeding demands

How to express breast milk

While your baby is not breastfeeding (during the first two to three days after diagnosis and any other time breastfeeding is interrupted such as illness), you will need to express milk to keep up your supply.

You can express your milk by hand or with a breast pump – using an electric or hand pump. You will need to express approximately every three hours to maintain a good supply of milk.

Please ask your dietitian for a Temple Street Children's University Hospital booklet – **'Information for Parents on Expressing Breast Milk'**. If you have any questions about breastfeeding, go to www.breastfeeding.ie.

Stopping breastfeeding

If you wish to stop breastfeeding, it is best to plan it with the metabolic dietitians and stop it over a two to three week period.

Formula feeding

If you choose not to, or are unable to breastfeed, standard infant formula is an appropriate alternative, along with a protein-free formula. Some babies will be started on a set amount of synthetic protein to provide essential amino acids. This will be discussed in more details in chapter 4. A measured amount of standard infant formula and a measured amount of synthetic protein (if given) are given by bottle first and then a protein-free formula to appetite.

A measured amount of standard infant formula +/- a measured amount of synthetic protein are given first and then a protein-free formula to appetite.

If your baby is hungry in between feeds you may give extra protein-free formula in addition to the measured amounts of standard infant formula +/- measured amount of synthetic protein.

Protein-free formula

Protein-free formula contains all the nutrients needed for growth, except protein. This will be prescribed for your baby.

The amount of protein-free formula and synthetic protein and breast milk or standard infant formula will need to be adjusted from time to time to provide the right amount of protein to meet your baby's needs and keep their blood levels in an acceptable range. Your dietitian and doctor will advise you on this as your baby grows.

Unless your dietitian advises you to do so, it is not necessary to add anything to your baby's formula.

Preparing formula (standard infant formula or protein-free formula)

- Boil fresh water
- Leave to cool for 30 minutes
- Clean surfaces and wash hands
- Pour the required volume of water into a sterile bottle
- Add the appropriate number of scoops using the scoop provided (1 scoop per 30ml/1oz)
- If your baby needs 45ml of formula, add 2 scoops of powder to 60ml/2oz of cooled boiled water and then pour out the extra formula until 45ml is left in the bottle
- Shake well
- Cool quickly under cold running water
- Check temperature
- Throw away unused feed after 2 hours

Synthetic protein formula

Some babies will be recommended to start on a set amount of a synthetic protein drink that contains essential amino acids. For more information, see chapter 4.

To store the formula

- Make up each bottle as required. It is safest to prepare a fresh feed each time you need one, and to give it to your baby straight away. This is because warm milk provides ideal conditions for bacteria to grow – especially at room temperature.
- If you need to prepare feeds in advance to use later, make up individual bottles, cool them quickly and place in the back of the fridge (5°C or below). Throw away any feed in the fridge that you have not used within 24 hours.

For additional information please ask your dietitian for the booklet ‘**How to prepare your baby’s bottle feed**’ produced by Safefood (www.safefood.eu) and the HSE (www.hse.ie).

Additional information for feeding your baby

Most babies, whether they are breastfed or bottlefed, need five or more feeds every 24 hours until they are four to five months old.

- Provided your baby is well, only the protein-free formula and the synthetic protein (if advised) and breastfeeds or standard infant formula are needed until they are 17 to 26 weeks of age.
- Babies should not be given herbal or medicinal teas or baby juice. Extra vitamins and minerals are not needed **except for vitamin D**.

Vitamin D and your baby

Whether you choose to breastfeed or formula feed, you should give your baby 5 micrograms (5µg) of vitamin D3 every day, for the first year of life. There are a number of suitable infant vitamin D3 products available to buy in Ireland. The number of drops or amount of liquid required daily is different for each product. Read the product instructions carefully and ask your pharmacist for advice if needed. **Only one dose per day should be given.**

Recording Your Babies Intake

Below is an example of a **guide** of a feeding/daily record plan which may be helpful for you to record your baby's feeding pattern.

For breastfed babies:

Feeds

Date.....

Time	Number of measured feed	Protein-free formula (measured)	Synthetic protein (measured)	Breastfeed (to appetite)
7am	Feed 1	30ml	10ml	Right side
10.30am	Feed 2	30ml	10ml	Left side
1.30pm	Feed 3	30ml	10ml	Right side
2.30pm				Left side
4pm	Feed 4	30ml	10ml	Right side
7pm	Feed 5	30ml	10ml	Left side
10.30pm	Feed 6	30ml	10ml	Right side
1.00am	Feed 7	30ml	10ml	Left side
4am	Feed 8	30ml	10ml	Right side
5 am				Left side

For formula-fed babies:

Feeds

Date.....

Time	Number of measured feed	Standard infant formula (measured) e.g. Aptamil infant milk	Synthetic protein (measured)	Protein-free formula (to appetite)
7am	Feed 1	20ml	10ml	60ml
10.30am	Feed 2	20ml	10ml	20ml
1.30pm	Feed 3	20ml	10ml	30ml
2.30pm				60ml
4pm	Feed 4	20ml	10ml	40ml
7pm	Feed 5	20ml	10ml	60ml
10.30pm	Feed 6	20ml	10ml	10ml
1.00am	Feed 7	20ml	10ml	30ml
4am	Feed 8	20ml	10ml	50ml
5 am				30ml

For breastfed babies:

Feeds

Date.....

[illegible]

For formula-fed babies:

Feeds

Date.....

Time	Number of measured feed	Standard infant formula (measured)	Synthetic protein (measured)	Protein-free formula (to appetite)

Chapter 4

Understanding the Synthetic Protein

Synthetic protein is a specially made protein that contains only the essential amino acids. Essential amino acids are amino acids that the body cannot make. Some children with Urea Cycle Disorders are recommended to take a synthetic protein. This is to ensure that your child gets enough of these essential amino acids to grow, build and repair tissues. If too much of this synthetic protein is given, the extra protein that the body does not use can be converted into ammonia. Therefore, your dietitian will recommend a **set amount of synthetic protein** per day for your child.

If recommended, the synthetic protein is an essential part of the treatment of Urea Cycle Disorders. If it is not taken as recommended, ammonia and amino acid levels, can be outside the acceptable range which can cause health and growth problems.

The amount of synthetic protein needed by your child will be calculated by your dietitian. This will change throughout your child's life depending on their age, growth, ammonia and amino acid levels.

What to do if your child is refusing or struggling with their synthetic protein?

- Don't panic.
- Contact the metabolic team to explore methods of getting your child to take their synthetic protein.
- There are a variety of synthetic protein options which may be more suitable to your child's taste.
- **Never mix** the synthetic protein with food. It is very important for your child to accept the taste of the synthetic protein in its true form.
- **Never force** your child to take the synthetic protein.

There is a variety of synthetic protein available in different forms and flavours to suit lifestyles and taste preferences. For example; powders, gels/pastes and ready made liquids. These are available on prescription through your local pharmacy. Your dietitian will help you find the right product for your child.

The synthetic protein must be spread throughout the day. This helps keep ammonia and amino acid levels within an acceptable range. If your child is struggling to take all of their synthetic protein, please inform your dietitian.

If you experience any problems with supply or quality of the synthetic protein, please contact the companies directly. Contact numbers for the companies will be provided.

Chapter 5

Dietary Management

At present the dietary treatment for Urea Cycle Disorders is a low protein diet. **This is a diet for life.** The diet prevents a build-up of excess ammonia in your child's blood, while at the same time providing enough protein for their normal growth and development. There are five main components of the diet:

1. Synthetic protein (see chapter 4)
2. Unwell management (see chapter 7)
3. Foods to avoid
4. Exchange foods
5. Free foods

The Urea Cycle Disorder diet must be low in protein and therefore, natural protein must be restricted. The diet is best described by a traffic light system.

RED STOP:

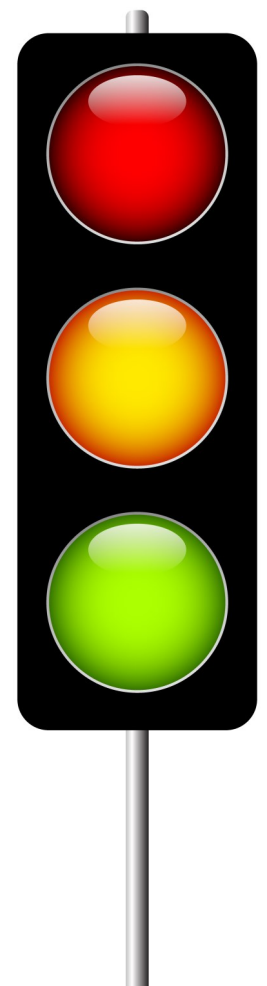
Do not eat these foods as they are too high in natural protein (small amounts may be allowed depending on tolerance).

ORANGE GO CAUTIOUSLY:

Foods that need to be counted as they contain some natural protein (exchange foods).

GREEN GO:

These foods contain very little natural protein and can be eaten freely.



GREEN GO: These foods can be eaten freely.

These foods are either naturally low in protein or have been specially made to be low in protein and so are 'free from exchanges' or do not have to be measured.

Some foods are naturally low in protein such as most fruit and vegetables, butter, margarine, cooking oils, jam, sugar and honey (**honey is not recommended for any infant under 12 months**). You can cook in all the usual ways (bake, grill, fry) and you can use herbs, spices and flavourings to add interest to foods.

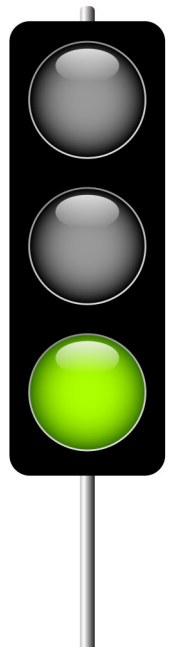
There is a large variety of specially manufactured low protein foods such as bread, pizza bases, pasta, rice, flour, cereals, milk, meat substitutes, cake mixes, biscuits and chocolate. These foods are available on prescription from your local pharmacy.

An up to date list of these foods will be given to you. All of the above foods should **make up the majority of your child's diet** and they do not need to be counted. There are many recipe books available using these low protein foods. Children with Urea Cycle Disorders are encouraged not to have too much sugar and fat for the same reasons as other children without Urea Cycle Disorders.

The following tables give examples of foods that are naturally low in protein and so are allowed freely in the diet.

Vegetables-fresh, tinned, frozen

Artichokes	Cucumber	Peppers
Asparagus	Curly kale	Pumpkin
Aubergine	Fennel	Radish
Beansprouts	French beans	Rocket
Beetroot	Garlic	Runner beans
Butternut squash	Gherkin	Samphire
Cabbage	Green beans	Scallions
Capers	Leeks	Shallots
Carrots	Lettuce	Spring onion
Cassava	Mangetout	Sugar snap peas
Celeriac	Okra	Swede
Celery	Onions	Tomato
Chayote	Pak choi	Turnip
Courgette	Parsnips	Water chestnuts



Fruit- fresh, tinned, frozen

Apple	Grapes	Passionfruit
Apricots	Guava	Peaches
Blackberries	Jackfruit	Pears
Blackcurrants	Kiwi	Physalis
Blueberries	Kumquat	Pineapple
Cherries	Lemons	Plums
Clementine	Limes	Prunes
Cranberries	Lychees	Raisins
Currants	Mandarins	Raspberries
Damsons	Mangoes	Rhubarb
Dates	Mangosteens	Satsumas
Dragon Fruit	Melon -cantaloupe	Star fruit
Figs	Melon -honeydew	Strawberries
Ginger	Nectarines	Sultanas
Gooseberries	Olives	Tangerines
Grapefruit	Papaya	Watermelon

For the following fruit and vegetables, if the portion identified below is taken **once a day**, this portion can be counted **freely**. However, if **more than this portion** is taken, any additional portions of this fruit or vegetable needs to be counted as **1 exchange**.

For example, 5 mushrooms in the day can be taken freely, but if 10 mushrooms are taken, then the additional 5 mushrooms should be counted as 1 exchange. Also, 80g of sweet potato in the day can be taken freely, but if 200g of sweet potato is taken, then the additional 120g should be counted as 1 ½ exchanges.

Fruit and vegetables to be taken in limited amounts

Bananas: 1 banana	Plaintain: 1 plaintain
Broccoli: 8 florets (85g)	Pomegranate: 1 pomegranate
Brussel sprouts: 6 sprouts (90g)	Sharon fruit: 1 sharon fruit
Cauliflower: 9 small florets (90g)	Spinach: 45g (1 blue scoop)
Mushrooms: 5 medium mushrooms (80g)	Sweet potato: 80g (1 ½ blue scoops)
Oranges: 1 orange	Yam: 100g = 1 exchange

Sugar	Sweets	Jams and preserves
Barley sugar	Any chewy sweets*	Chutneys
Brown	Boiled sweets	Golden syrup
Castor	Candyfloss	Honey
Glucose	Iced lollies	Jam-any type
Icing sugar	Lollipops	Marmalade
Jam sugar	Sorbet	Maple syrup
White	Sherbet	Treacle
	Suitable jellies*	
	Other suitable sweets from treats list	

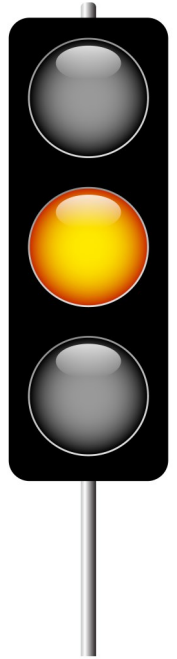
*always check the foods listed above for gelatine

Seasonings	Miscellaneous	Food essences and colourings
Chilli	Baking powder	Almond essence
Curry paste	Bicarbonate of soda	Cochineal
Curry powder	Buttercream icing	Peppermint
French dressing	Cornflour	Vanilla
Herbs	Cream of tartar	
Mint sauce and jelly	Sprinkles	
Mustard	Some custard powders	
Pepper	Pasta, noodles or rice made from	
Salt	konjac (Asian root vegetable)	
Spices	Sago	
Vinegar	Tapioca	

Fats and oils	Drinks	Sauces
Butter	Some coconut milks	Apple sauce
Coconut oil	Pure fruit juice	Balsamic vinegar
Duck Fat	Hemp milk	Cranberry sauce
Goose fat	High energy drinks	Chutney
Fry light spray	Minerals	Horseradish sauce
Grape seed oil	Rice milk (not suitable in children under 5 years or pregnant/ lactating women)	Ketchup, brown sauce
Garlic oil	Squashes/cordial	Most mayonnaises, salad creams
Lard	Tea/coffee/herbal tea	Mustard
Margarine	Water/ soda water/mineral water	Pasta sauces, cook- in sauces
Olive oil		Piri Piri sauce
Rapeseed oil		Relish
Sunflower oil		Salad dressing
Sesame oil		Soya sauce (check label)
Suitable low fat spread	See chapter 13 for suitable alcoholic drinks	Sweet chilli sauce
Vegetable oil		Tomato salsa
		Tabasco sauce
		Worcestershire sauce

ORANGE GO CAUTIOUSLY: These foods can be eaten in measured amounts and are known as exchanges.

Your child requires a small amount of protein for growth, development and repair. Foods such as cereals, rice, beans, peas and potatoes contain a small amount of protein. These are given in small **measured quantities** called exchanges. Every child will have a set number of exchanges per day. The number of exchanges your child is allowed will change depending on their blood ammonia and amino acid levels. This is often referred to as natural protein. Your dietitian will educate you on counting exchanges and give you a booklet containing commonly used exchange foods.



An exchange is the amount of any food that contains 1 gram of protein.

1 exchange = 1 gram (g) protein

Examples of foods that contain 1 exchange include:

- 1 ½ level blue scoops of mashed potato
- 2 tablespoons of peas
- 1 level blue scoop of sweetcorn
- 1 blue scoop of boiled rice
- 1 tablespoon of baked beans
- 1 tablespoon of chickpeas

These foods are spread out between the day's meals to provide small amounts of essential protein. It is important to be able to read food labels so that you can count exchanges for your child. This will be discussed in detail later in this chapter.

RED STOP: Do not eat these foods (small amounts may be allowed depending on tolerance)

These foods are high in protein and so are not allowed.

Meat: Beef, lamb, pork, ham, bacon, chicken, turkey, duck and game.

Offal: Liver, tongue and kidney.

Processed meat: Sausages, rashers, pudding, corned beef, chorizo, meat products e.g. burgers, meat paste, meat pate and meat pies.

Fish: All kinds including shellfish, frozen or tinned. Also processed fish e.g. fish fingers and fish cakes.

Eggs: All varieties.

Milk and milk products: Milk (including cows, goats, soya), yogurt, yogurt drinks, cream, ice cream and chocolate.

Cheese: All varieties (*some vegan cheese and cheese spreads may be suitable*).

Soya: Foods made from soya such as TVP, Quorn and Tofu.

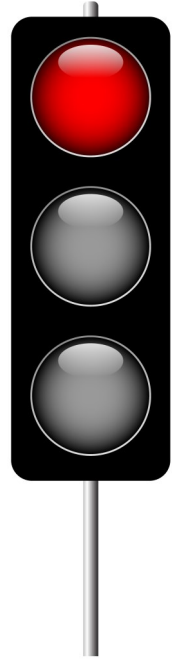
Vegetarian products: Vegetarian meals, burgers and sausages.

Nuts and seeds: Including peanut butter and nut spreads.

Bread: White and brown bread, soda, pitta, naan, bagels, ciabatta, wraps, croissants and scones.

Flour based foods: Pasta, cakes and biscuits.

Gelatine: Jelly, jellies and sweets containing gelatine.



Remember:
For every food that your child cannot have, there are suitable low protein alternatives available.

Label Reading

Reading food labels plays an important role in helping you decide whether a food is suitable or not. Reading food labels can be confusing but you will have support from the dietitian to help you with this. Once you know how to read food labels, this can increase the variety of foods in your child's diet. It is important to check labels regularly as ingredients and protein content can change.

Tips on Reading Food Labels:

- Firstly look at the ingredients to see what the product contains.
- Sometimes, the nutritional information states that some protein is present within the product. However, if you look at the ingredients you may see that all the ingredients present in that product are foods allowed freely. In this situation, the product can be given freely e.g. tinned tomatoes (4g of protein in a can of tinned tomatoes according to the nutritional information). *This product is not counted because tomato is a free food.*
- If the product contains exchange containing foods e.g. rice, potato, peas or cream, then the protein should be counted as described in the nutrition label. For example, a jar of curry sauce contains cream, therefore it must be counted.
- Another example of this would be a soup which contains peas. This product contains peas which is an exchange food; therefore, the protein content needs to be counted as described on the label. For example, if there is 1.0g of protein per carton of soup then this product needs to be counted as 1 exchange.

Some foods or drinks will already have the protein content per portion written on the food label. Use the guide below to help you.

Protein content per portion	Number of Exchanges
0 – 0.3g	Free
0.4g – 0.7g	$\frac{1}{2}$
0.8g – 1.2g	1
1.3g – 1.7g	$1 \frac{1}{2}$
1.8g – 2.2g	2

However, this is not always the case. Sometimes, you will have to calculate the protein content per portion yourself.

To do this, you will need to know two things:

1. The weight/amount of the food to be eaten
2. The protein content per 100g of the food

This is how it is done:

$$\frac{\text{Weight of food to be eaten} \times \text{Protein content per 100g}}{100}$$

Example 1: Tinned Ratatouille Provençale

390g carton

Ingredients: Tomato, Courgette, Tomato Purée, Aubergine, Peppers, Onion, Vegetable Oil, Sugar, Water, Salt, Pepper Extract, Garlic Extract, Thyme Extract, Lemon Concentrate

Nutritional Information

Typical Values	Per 100g	Per 195g (½ carton)
Energy (kJ)	179	350
Energy (kcal)	43	85
Fat	2.0	3.9
(of which saturates)	0.3	0.6
Carbohydrate	4.2	8.2
(of which sugars)	3.0	5.9
Fibre	1.9	3.7
Protein	1.1	2.1
Salt	0.5	1.0

Step 1: Look at the ingredients. All the ingredients are 'free' foods so you do not need to use the nutritional label to check the protein content.

Example 2: Tomato Pasta Sauce

150g carton

Ingredients: Tomatoes, **Cream**, Lemon Juice, **Mascarpone Cheese**, Cornflour, Sugar, Tomato Paste, Salt, Fat Powder, Basil, Garlic, Spices, Herbs

Nutritional Information

Typical Values	Per 100g	Per 150g carton
Energy (kJ)	237	356
Energy (kcal)	57	86
Fat	2.6	3.9
(of which saturates)	1.7	2.6
Carbohydrate	6.6	9.9
(of which sugars)	3.8	5.7
Fibre	1.9	2.9
Protein	1.2	1.8
Salt	0.55	0.83

Step 1: Look at the ingredients. **Cream** and **mascarpone cheese** are both 'exchange'/protein containing foods. Therefore, you need to use the nutritional label to work out the protein content.

Step 2: Look at the nutritional label

1 carton = 1.8g protein

Therefore, 1 carton = 2 exchanges.

Example 3: Garden Vegetable Soup

400g tub

Ingredients: Water, Carrot, Turnip, Onion, Celery, **Potato**, Parsnip, Leek, **Sweetcorn**, **Red Lentils** (2%), **Peas**, Vegetable Stock (Salt, Onions, Celery, Carrots, Turmeric, Parsley), Salt, Tomato Purée, Black Pepper

Nutritional Information

Typical Values	Per 100g
Energy (kJ)	105
Energy (kcal)	25
Fat	0.2
(of which saturates)	<0.1
Carbohydrate	7.3
(of which sugars)	2.1
Fibre	1.7
Protein	1.6
Salt	0.6

Step 1: Look at the ingredients. **Potatoes**, **sweetcorn**, **red lentils** and **peas** are all exchange foods. Therefore, you need to use the nutritional label to work out the protein content.

Step 2: Look at the nutritional label

Protein per 100g = 1.6g protein. However, $\frac{1}{2}$ tub = 200g

Step 3: Use the formula to work out how many exchanges in the $\frac{1}{2}$ tub.

Protein content per 100g (1.6g) x Weight of product to be eaten ($\frac{1}{2}$ tub = 200g)

100

200g ($\frac{1}{2}$ tub) = 3.2g protein

Therefore, $\frac{1}{2}$ tub = 3 exchanges.

Example 4: Rice Cakes

Ingredients: Wholegrain Rice (100 %)

Nutritional Information

Typical Values	Per 100g	Per cake (6.7g)
Energy (kJ)	1639	110
Energy (kcal)	387	26
Fat	3.7	0.2
(of which saturates)	0.7	<0.1
Carbohydrate	78	5.2
(of which sugars)	0.7	0.1
Fibre	3.4	0.2
Protein	8.5	0.6
Salt	0.01	<0.01

Step 1: Look at the ingredients. Rice is an exchange food. Therefore, you need to use the nutritional label to work out the protein content.

Step 2: Look at the nutritional label

1 cake = 0.6g protein = $\frac{1}{2}$ exchange

2 cakes = 1.2g protein = 1 exchange

Additional Information on Reading Food Labels

For foods that are not on our exchange lists, you may need to work out the amount of food that contains 1 exchange e.g. breakfast cereals

- Weight of Product that is 1 Exchange:

$$\frac{1 \times 100}{\text{Protein content per 100g}}$$

- Weight of Product For Your Required Number of Exchanges:

$$\frac{\text{No. of Exchanges} \times 100}{\text{Protein content per 100g}}$$

The following example shows you how to use this information.

Example 5: Oat Flakes

Ingredients: Wholegrain **Oats** (84%), Maltodextrin, Sugar, Malted **Barley** Extract, Potassium Chloride, Salt, Niacin, Iron, Pantothenic Acid (B5), Thiamin (B1), Riboflavin (B2), Vitamin B6, Folic Acid, Vitamin B12.

Nutritional Information

Typical Values	Per 100g
Energy (kJ)	1618
Energy (kcal)	387
Fat	5.5
(of which saturates)	1.1
Carbohydrate	73
(of which sugars)	1.3
Fibre	6.1
Protein	11.1
Salt	trace

Step 1: Look at the ingredients. Oats are an exchange food. Therefore, you need to use the nutritional label to work out the protein content.

Step 2: Look at the nutritional label

Protein per 100g = 11.1g protein

Step 3: Use the formula to work out how many grams of these oat flakes are 1 exchange.

1 x 100

11.1 = 9g of oatflakes for 1 exchange

2 x 100

11.1 = 18g of oatflakes for 2 exchanges

You would need to weigh out this product.

Preparing a low protein meal & a family meal at the same time

Below are some tips on how to make the low protein meal similar to family meals. It is important that your child feels included in the family meal. They should eat at the same time as the rest of the family. Try to include them in the cooking as often as possible.

Family Meal	Low Protein Meal
Roast meat with roast vegetables and gravy	Roast vegetables served with: <ul style="list-style-type: none"> • Gravy and potato (count exchanges) • Low protein stuffing • Low protein burger/ sausage mix (count exchanges) • Low protein Yorkshire pudding
Quiche or Pie	Grated vegetables (courgette, parsnip, carrot, butternut squash, sweet potato (may need to count exchanges)): <ul style="list-style-type: none"> • Baked as a pie with herbs and white lasagne sauce using low protein pastry • With pasta sauce and a low protein bread crumb topping • Piled into a vol-au-vent shell (count exchanges)
Grilled meat and steamed vegetables/ salad	Fried or baked aubergine or courgette in low protein breadcrumbs with steamed vegetables/salad
Stir fry with rice or noodles <i>(cook meat separately and use the vegetable stir fry for everyone)</i>	Stir fry vegetables with suitable soy sauce, ginger and garlic or use a suitable stir-fry sauce Cook low protein spaghetti and toss into stir fried vegetables to resemble noodles Use low protein flat noodles, konjac noodles, rice noodles (count exchanges if applicable)

Family Meal	Low Protein Meal
Sausage, tomato sauce, mash or chips and vegetables	Use sausage/burger mix and make into sausage shape (count exchanges) with potatoes (count exchanges) or sweet potato chips (may need to count exchanges) or parsnip chips
Burger and chips	Use burger mix to make a burger shape (count exchanges) and serve between a toasted low protein roll with salad and sweet potato or potato chips/wedges (count exchanges if applicable)
Macaroni Cheese	Use white lasagne sauce, herbs and low protein macaroni pasta and vegan cheese
Pizza	Low protein bread or pizza base – spread with tomato puree and top with vegetables and low protein cheese (count exchanges if applicable)
Barbeque	Vegetable skewers and baked potato (count exchanges) with coleslaw. Or make up burger mix (count exchanges) to a burger shape and serve in a low protein roll with coleslaw
Spaghetti Bolognese	Low protein spaghetti, fry off desired vegetables and tomato based bolognese sauce. Use burger mix to make meatballs if desired (count exchanges)
Curry	Use a curry paste with tinned tomatoes and tomato paste for flavour and add to fried vegetables of choice. Add a small amount of coconut milk if desired (may need to count as exchanges). Serve with low protein rice



Chapter 6

Monitoring

Monitoring blood ammonia and amino acids is an important part of managing Urea Cycle Disorders. Keeping blood ammonia and amino acid levels under control reduces the risk of problems.

A Urea Cycle Disorder requires frequent blood samples to monitor the effectiveness of treatment. The results of these tests are used to adjust the diet and medications as required. Additional bloods may be required depending on blood control and other factors such as illness, surgical procedures and pregnancy. The metabolic team will decide on this and inform you what is necessary at a given time.

Blood samples need to be taken in your local hospital. Bloods for **amino acids** and **ammonia** are the routine requests for Urea Cycle Disorders. Ammonia may be checked in your local hospital (depending on the hospital). The other bloods will be sent to the Metabolic Laboratory in Temple Street for analysis. You will be given the necessary request forms for these bloods at your clinic appointments.

Remember if the procedure of taking the blood sample is traumatic, this can cause the ammonia results to be higher than it actually is in the blood.

If ammonia levels or levels of certain amino acids are too high or too low, your metabolic doctor (or dietitian) will advise you appropriately on changes to make to the diet and medications. A number of factors cause blood levels to increase or decrease outside of the target range.

Ammonia levels and certain amino acids levels may increase when:

- Natural protein intake (exchanges) is too high
- Intake of synthetic protein is too high (if applicable)
- Alternatively, intake of protein (natural and synthetic) and therefore essential amino acids is too low causing the body to break down its own muscle tissue for nutrients. This releases protein into blood and this leads to increased ammonia levels
- A child's rate of growth has slowed meaning less protein is being used to build new proteins
- Illness, vaccinations and teething may make it difficult to eat or take the synthetic protein or high calorie drink. This can cause high ammonia levels from the breakdown of the body's own muscle tissue

Ammonia levels and certain amino acids levels may increase when continued:

- Insufficient calorie intake results in the body breaking down its own muscle tissue for calories and causes the release of protein into the blood, resulting in a build-up of ammonia
- Adjustments to the medications are needed

Ammonia levels and certain amino acid levels may decrease when:

- Intake of natural protein (exchanges) is lower than recommended
- Intake of synthetic protein is too low (if applicable)
- Growth in a child causes levels to fall as protein is used to build new body tissues

It is really important to inform your metabolic dietitian or doctor of any issues that may have arisen in your child during the days prior to levels being taken e.g. overexchanging, missed medications, etc.

Frequency of monitoring blood levels

Frequency of blood monitoring depends on the individual's age, condition, ammonia and amino acid levels. This is determined by the metabolic team.

Results

The metabolic doctor will contact you with the results of your child's blood results. The contact information for the metabolic doctors is as follows:

01-8784200 and ask to speak to the metabolic registrar on-call.



In case of emergency, please contact the metabolic nurse or doctor on-call via switch 01-8784200.



Chapter 7

Illness

Children with Urea Cycle Disorders will have the same number of coughs, colds and other illnesses as all children, but they need extra care because of the risk of rapidly increasing ammonia levels during periods of illness. **They can become very sick, very quickly. When unwell, it is essential to contact the metabolic team.** The contact number is 01 8784200 and ask to speak to the metabolic registrar on-call between 9am and 9pm. After these hours, ask to speak to the metabolic nurse on-call.

If you have any concerns, ring the emergency number you have been given. It is better to be overcautious.

During illness, the body starts to break down its own body tissue, releasing protein into the blood stream. Levels of ammonia increase quickly which can make your child very ill. If left untreated it can become life threatening.

During illness, high ammonia levels can be reduced by giving appropriate dietary management and high calorie drinks. When unwell, the natural protein will be either reduced or stopped.

Synthetic protein may also be stopped, as when unwell, it can contribute to high ammonia levels. All persons with Urea Cycle Disorders will have an individualised 'unwell plan'. This unwell plan will be updated regularly by the dietitian when you attend your outpatient clinic appointment. This unwell plan will guide you on the different unwell feeds needed and the volume required daily when unwell. Stop natural protein and synthetic protein and commence the unwell plan until you have spoken to the metabolic team. The metabolic team will advise you on how much natural protein and synthetic protein to take and if additional medications are required.

If a child is not drinking enough of their unwell feeds, it can be given through a nasogastric tube (a tube that passes through the nose and into the stomach). You may be trained how to insert this tube so it can be done at home. Alternatively, you will need to go to your local hospital. If your child is not tolerating feeds through the nasogastric tube, intravenous feeds (IV drip feeds) will be used; this can only be done in hospital.

Call the 24 hour emergency numbers you have been given immediately if your child has ANY of the following:



- Vomiting
- Diarrhoea (very loose or watery stools)
- Refusing feeds
- Poor appetite for food
- Temperature over 38°C
- Excessive sleepiness
- Low energy levels
- Slurred speech
- Unsteadiness or feeling wobbly
- Irritability

When unwell, it will be necessary to do more regular bloods. Any further deterioration in your child's condition should be immediately reported to the metabolic team. The metabolic team need to be contacted even if your child has previously coped with infections and illness. Sometimes, a relatively mild illness may require an admission to hospital even when you have coped with their illness at home before.

An admission to hospital may also be required at the time of vaccinations, surgical procedures e.g. dental extractions, or injuries e.g. broken limb.

If your child needs to be admitted to your local hospital, please ensure you bring a copy of the unwell plan, emergency medical letter, adequate supplies of all products that you need for the unwell feeds and medications.

At home, make sure you have:

- Phone numbers for your metabolic team and local doctor
- Copies of your unwell plan stored in an accessible place
- In date supplies of all products needed for the unwell plan
- Sufficient supplies available for a number of days
- Emergency letter from your metabolic doctor outlining the Urea Cycle Disorder and its management during illness
- Supply of medications

During illness, your child's medications are likely to be increased. You will be guided by the metabolic team on this. As your child begins to recover, their natural protein and synthetic protein intake will gradually be increased; you will be guided by the metabolic team regarding this.

You may also need to use your unwell plan if ammonia levels are high for unknown reasons.

Dialysis

If levels are very high during the hospital stay, dialysis may be used to remove very high levels of ammonia.

Vaccinations

Vaccinations are a way to protect your child from serious illnesses. Routine vaccinations should be given according to the national immunisation scheme www.immunisation.ie. Your metabolic team will discuss this with you and whether additional vaccinations would be beneficial to your child such as flu vaccinations. Your child will need to go onto their unwell plan for vaccinations and during their early years may need to be admitted to hospital for monitoring around this time. Extra medications may also be needed.

Dietary plans

Each child will have their own individual dietary plan. The plan will provide you with information needed to manage your child's diet **when well and unwell**. It will be updated by the dietitian after your child's outpatient appointment to provide an up to date summary of the diet and feeds you use. The dietary plan outlines the feeds required daily, recipes for these feeds and other dietary products needed. The dietary plan consists of two to three pages. It is important that you understand this plan and you keep it in an accessible location. It may be useful to keep several copies of the unwell plan. When you get a new plan, ensure you dispose of the old plan.

Page 1: The Well Plan

The first page of the plan is the well plan. This gives you all the information on the number of exchanges required and feeds that your child needs to be take daily when well. It looks like this:

PAGE 1

Patient Name

PART A This section contains your personal information and is **for hospital use only**.

Diagnosis:	
Date:	
Current weight:	_____ Kg

PART B This section contains information on the diet and feeds when **well**. This information has been agreed with your consultant and is **for use by the doctors and dietitians**.

DIETARY AIMS WHEN WELL	
Energy	_____ kcal per day (_____ kcal/kg)
Natural Protein	_____ g (_____ g/kg)
Synthetic Protein	_____ g (_____ g/kg) or mls per day
Total Protein	_____ g (_____ g/kg)
Fluid requirements	_____ ml per day
Vitamins & Minerals	
AA/DHA supplement	
High Calorie Drinks	_____ per day
Remainder of kcal to be achieved from exchanges and low protein foods.	

PART C This is the **most important section for you**. It contains information on the exchanges and the volumes of each feed and other dietary products that needs to be taken on a daily basis when **well**. Your child's exchanges may change between outpatient appointments. It is important to follow the advice you are given between appointments.

DIETARY PLAN WHEN WELL	
Natural Protein	_____ exchanges or ml
Synthetic Protein	_____ ml per day
High Calorie Drinks	_____ minimum ml
Vitamins & Minerals	
AA/DHA supplement	

PART D: This section is also important for you. It contains the recipes required when well. These feeds will need to be made up daily when **well**. These have been put together by your dietitian.

Well Recipes

Synthetic Protein	<u>Per 100ml</u>
Natural Protein	<u>Per 100ml</u>
High Calorie Drink	<u>Per 100ml</u>

Page 2: The Unwell Plan

If sick, the Unwell Plan must be followed. Page two on your child's dietary plan provides information on your child's unwell plan including recipes for unwell feeds and the volumes of these feeds that need to be taken when unwell. It is important that if unwell at home the natural and synthetic protein is stopped, start the unwell plan and contact the metabolic team. A hospital admission may be required if the unwell plan cannot be achieved.

PAGE 2

PART E This contains information on the diet and feeds when **unwell**. This information has been agreed with your consultant and is for **use by the doctors and dietitians**.

DIETARY AIMS WHEN UNWELL	
Energy	_____kcal/day (_____kcal/kg)
Natural Protein	As per metabolic team.
Synthetic Protein	May be stopped or reduced-advice will be given by the metabolic team.
Vitamins & Minerals	
AA/DHA supplement	
High Calorie Drink (CHO/Fat)	See plan below

PART F This section shows the volumes of all **unwell feeds** when unwell. If your child is unwell, you need to contact the metabolic team for advice. They will advise you on the number of exchanges (natural protein) and the amount of synthetic protein (**may be stopped or reduced**) to take daily. You must ensure an adequate volume of the high calorie drink is taken to ensure calorie needs are met. The volume of the high calorie drink needed will depend on the number of exchanges that are recommended by the metabolic team. These may change daily when your child is unwell.

DIETARY PLAN WHEN UNWELL	
Natural Protein	Exchanges are stopped or reduced as directed by the Metabolic Team.
Synthetic Protein	May be stopped or reduced-advice will be given by the metabolic team.
Exchanges	
If Reduced to Ex	Give _____ml High Calorie Drink (___ml x feeds)
If Reduced to Ex	Give _____ml High Calorie Drink (___ml x feeds)
If Reduced on Ex	Give _____ml High Calorie Drink (___ml x feeds)

PART G These are your unwell plan recipes. They may be different to your well plan recipes so please make sure that you understand them and make them up correctly.

Unwell Recipes

Synthetic Protein	<u>Per 100ml</u>
Natural Protein	<u>Per 100ml</u>
High Calorie Feed	<u>Per 100ml</u>

Page 3: The Unwell Plan

Sometimes a third page will be added.

This will give a list of protein free foods (both naturally protein free and low protein manufactured foods) that can be taken during a period of illness. Depending on the food taken, the volume of the high calorie drink needed can be reduced.; for example, if your child has one slice of low protein bread, the volume of the high calorie drink required may be reduced. The dietitian will discuss this with you in more detail.

Chapter 8

Weaning

Weaning is an exciting time in your baby's life when you begin to introduce some solid foods. It is recommended that weaning begins no earlier than 17 weeks (4 months) of age. This is because your baby's tummy is not ready for solid food before this. We recommend not to delay weaning beyond 26 weeks of age as there is a 'window of opportunity' to allow your baby to develop the skills to eat and accept new tastes and textures. It will also ensure your baby eats a balanced diet. Protein 'free' foods are started first.

Your dietitian will give you a copy of '**Weaning your baby on a Urea Cycle Disorder diet**' before the time comes to wean and will discuss weaning in detail with you.

This booklet can also be downloaded from:

<http://metabolic.ie/patient-family-information/metabolic-conditions/urea-cycle-disorders/>



Chapter 9

Preparing for Childcare, Pre-School & School

Preplanning

Children with Urea Cycle Disorders can easily make the transition to childcare and school with preparation. There is no reason why their condition cannot remain well controlled. There needs to be ongoing support and communication between the family, school and the metabolic team.

The following suggestions are a guide for planning and discussing your child's Urea Cycle Disorder with the childcare or school.

Discussing Urea Cycle Disorders with childcare and school

Who do you need to inform?

It is important that teachers and others at your child's childcare or school understand why your child needs a special diet, why it needs careful supervision, why they require medications and also that they know about particular concerns with illness.

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

- the principal and admissions staff (when you are planning your child's enrolment)
- the class teacher
- the supervisor of after school care
- the tuck-shop or canteen staff

What to discuss with staff

Firstly, they will need a basic understanding of Urea Cycle Disorders and the importance of the low protein diet, medications and any particular unwell management.

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 inherited and non-contagious.
- Children with this condition cannot break down protein in food.
- All children need a certain amount of protein for growth and repair of the body, but in your child's condition the extra protein can cause problems.
- Eating the wrong foods will not make them immediately sick, but may have a detrimental effect over the day or longer-term.
- Your child is on a very specific diet that is calculated by a specialist metabolic team, with portions/exchanges measured out daily by the child's family – so it is 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 the child has eaten food that is not allowed or does not eat foods that are sent from home.

The school must inform you if your child is unwell or has an accident, because urgent treatment may be needed to prevent more serious problems occurring.

You can request a letter or photocopy relevant sections of this handbook for the childcare or school to refer to as needed.

Food at school

Each childcare facility or school will vary in what they provide in terms of meals or canteen food.

You will need to decide whether to:

- provide all the food your child will eat at school
- provide the main meals but use the school canteen or preschool meals for snacks, such as fruit or salad plates – you could also give the school a list of foods that are 'free' (contain minimal or no protein) or a list of exchange foods (foods allowed in measurable quantities)
- use only the school facilities – by either pre-ordering the selected foods from the menu, or providing low protein food for the school canteen to make into meals

Taking synthetic protein or calorie drink in school

Your child is likely to be happy to take the synthetic protein or calorie drink at the usual times at school. Label it with their name and store it in the fridge or at room temperature depending on their preference. Encourage your child to take the synthetic protein or calorie drink to school to help spread it throughout the day.

Parties and cooking days

Prepare for these ahead of time by keeping a supply of pre-made food which has the protein measured or low protein treats in the freezer.

If the class is learning about food or cooking, your child can share food using the low protein equivalents, so they are not excluded. Other children can also sample the food.

What to discuss with your child

Talking to your child is an important part of preparing for childcare and 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 free and which to count
- bringing home uneaten food in the lunch box so that you can calculate exchanges
- buying only suitable food from the canteen
- not swapping lunches with friends
- deciding when to drink the synthetic protein or calorie drink
- how to explain the different diet to other children
- what to do if being teased

Offer encouragement to your child by:

- giving positive messages about the foods they can have
- discussing this way of eating keeps them healthy and helps them to grow
- reassuring them that other people are on a special diet so your child knows they are not the only one



Letters for childcare/school are available from the metabolic team or on our website www.metabolic.ie

Chapter 10

Encouraging Independence

When your child is very young, you have the main responsibility for managing their condition. Treat your child like other children and try not to be over sympathetic or protective. Never say anything negative about the food, synthetic protein or medications to your child. While managing their Urea Cycle Disorder will be part of both you and your child's life, it will be just that: a part of life. A positive and an encouraging attitude by all the family is much more likely to lead to their acceptance of the treatment as they grow.

It is important to start sharing this responsibility with your child as they grow older. Looking after their diet and medications needs to be incorporated into their care, as much as learning to wash their hands after using the toilet or tidying up their toys.

Fostering independence in managing their Urea Cycle Disorder has a number of advantages. It:

- Encourages **acceptance**.
- Helps them develop a good **understanding** of Urea Cycle Disorders.
- Increases their **confidence** in controlling their condition.
- Helps you and your child **work together** better as a team.

By allowing your child to take charge as they mature, some of the more difficult aspects of their treatment become a shared responsibility. When children with Urea Cycle Disorders are able to accept and take some responsibility for managing their condition in everyday life, their long-term adjustment and dietary control tend to be better. This helps them prepare for adult life when they will need to take full responsibility for their Urea Cycle Disorder. As your child grows, your role will change from managing the diet and medications to supervising it. Later, you will observe and support your child as *they* make the important decisions.

What to expect of your child at different stages

Toddlers: 2–5 years

- Are aware that they have a special diet
- Know they need to take their synthetic protein and/or calorie drink (if applicable)
- Know they need to take their medications
- Know they need to have blood tests
- Know to check new foods with their parents



Toddlers: 2–5 years continued

- Watch you prepare the synthetic protein and/or calorie drink
- Start to learn about yes/no foods
- Are aware of exchange foods that need to be counted
- Know they have to take special drinks if they are sick and may have to go to hospital

Early childhood: 5–8 years

- Understand that they have a condition called a Urea Cycle Disorder (or specific name)
- Understand that they cannot eat high protein foods
- Have a basic knowledge of why the synthetic protein and calorie drink (if applicable) are important
- Help you prepare the synthetic protein and/or calorie drink
- Know that their medications are important
- Know what is considered a high ammonia level
- Practice using scoops to measure foods and count exchanges
- Have ideas about meal choices
- Learn to select low protein foods with assistance in social situations
- Know that they have to go to hospital regularly to have their blood tests done
- Know that they have to take special drinks when they are sick and may need to go to hospital



Late childhood: 8–12 years

- Have a basic understanding of what is a Urea Cycle Disorder and its specific name, protein, amino acids, ammonia, their diet and medications
- Are able to prepare their own synthetic protein and/or calorie drink
- Understand that taking synthetic protein and/or calorie drink helps provide nutrition and helps ensure good blood levels
- Can identify the different medications and understand that taking these medications when required is important
- Can measure foods accurately and count exchanges
- Can read food labels with assistance
- Are able to make appropriate meal choices
- Know how often they need to go to hospital to have their blood tests carried out.
- Know that they have to take special drinks when they are sick and may need to go to hospital to have a tube inserted or a drip put in. They also need to be able to recall the recent intake of these special drinks

Early teens: 12–14 years

- Have an understanding of what is a Urea Cycle Disorder and its specific name, protein, amino acids, ammonia, the effects of illness and medications
- Understand the risks and effects of high blood amino acids and ammonia levels on their health
- Prepare their own synthetic protein and/or calorie drink all the time
- Prepare their own medications and take at the correct times with assistance
- Make an effort to expand low protein diet choices
- Are able to make appropriate meal choices and prepare some basic meals for themselves
- Are able to keep a food diary to record their protein intake
- Read and calculate exchanges from product labels without assistance
- Remember when they need to go to hospital to have their blood tests carried out
- Know the recommended levels for blood ammonia
- Are able to engage in clinic appointments
- Know how to follow their dietary unwell plan with assistance and how to contact the hospital if they are unwell
- Know that they will need to go to hospital if they are unable to tolerate their dietary unwell plan or if their ammonia levels are high
- Know that adjustments to medications may need to be made if they are unwell
- For girls: Are aware that pregnancy should be planned with Urea Cycle Disorders

Late teens: 14–18 years

- Have a thorough understanding of their specific Urea Cycle Disorder and its effect on their body
- Prepare their own synthetic protein and/or calorie drink
- Can measure exchanges accurately
- Are able to make and prepare appropriate meal choices
- Remember to go to their local hospital independently for blood samples and to take phone calls with their parents from the metabolic dietitian or doctor ringing with their level results
- Are able to make their own dietary changes depending on the exchanges recommended
- Are able to make changes to medications depending on recommendations. Able to take the correct medications at the correct times without assistance
- Know how often clinic visits are needed and take responsibility for making their own appointments
- Are able to engage in clinic appointments

Late teens: 14–18 years continued

- Know how to contact their metabolic team when required
- Know that they will need to go to hospital if they are unable to tolerate their dietary unwell plan or if their ammonia levels are high
- Know how to follow their dietary unwell plan and how to contact the hospital if they are unwell
- Know how to make adjustments to medications when unwell
- For girls: Are aware that pregnancy should be planned with Urea Cycle Disorders



Talking about Urea Cycle Disorders with your child

Talking to your child about their Urea Cycle Disorder 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 synthetic protein and/or calorie drink to school, or if to take food to a party or sleepover, or choose food they are allowed from what is provided. They also need to know the importance of taking their medications.

Having a Urea Cycle Disorder, they will be asked questions about their diet.

Frequently asked questions include:	Here are some possible answers:
Why do you not eat meat?	I am on a special diet. I do not mind not eating meat – I have never had it and do not miss it.
Are you vegetarian?	My diet is like a vegan diet but even stricter.
Are you sick?	No, I am not sick – I am healthy and my diet keeps me healthy.
Is it contagious?	It is no more contagious than being a vegetarian. You cannot catch a Urea Cycle Disorder, you have to be born with it.
How can you drink your synthetic protein/calorie drink?	I have taken a synthetic protein since I was a baby, so I'm used to it. I am used to having synthetic protein/calorie drink. It is like medicine – I need it to be healthy.
Why did you have to go to the hospital when you were sick?	Sometimes when I get sick, I need extra care.
Why do you have to take those supplements?	The supplements keep me healthy.

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

Games

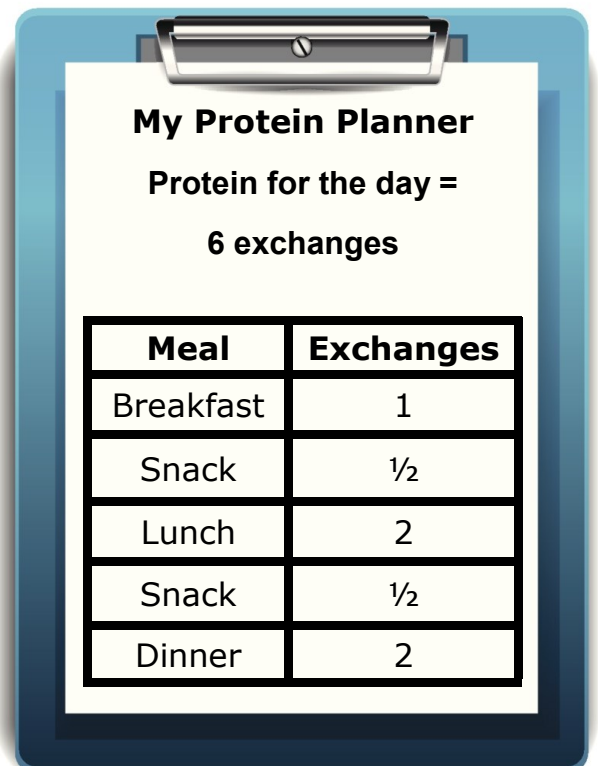
Regular discussions about Urea Cycle Disorders with your child will help them feel more comfortable talking about their Urea Cycle Disorder and give them the confidence to ask you questions and share their concerns. You can help your child to understand their Urea Cycle Disorder in a variety of ways 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 Urea Cycle Disorder Scrapbook.
- Letting your child rate new foods or meals from 1–5 to encourage them to try new foods.
- Involve your child in cooking low protein recipes to take to school or share with the family. Children as young as 2 or 3 years of age can help you pour ingredients into a bowl and stir. School age children can help peel and cut vegetables and measure ingredients. As children get older they can plan a meal and follow a simple recipe.
- Trying different ways of taking their dietary unwell plan when they are well and rating them from 1 to 5.
- A star chart/reward system for drinking their synthetic protein/calorie drink, managing their diet, taking their medications and doing their blood tests.

The more the Urea Cycle Disorder can become just another part of life, the easier their life and yours will be.

Daily exchange planner

Why not count exchanges together? Attach a magnetic board to the wall and write in the amount of exchanges you will offer at each meal. For each exchange, place a magnet on the board. As the exchanges are eaten during the day, your child removes 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. If your child is on limited exchanges it would be helpful to use different coloured magnets to indicate 1 exchange and $\frac{1}{2}$ exchange as some foods may be measured in $\frac{1}{2}$ exchanges. Beads on a string can also be used to demonstrate counting exchanges.



My Protein Planner	
Protein for the day =	
6 exchanges	
Meal	Exchanges
Breakfast	1
Snack	$\frac{1}{2}$
Lunch	2
Snack	$\frac{1}{2}$
Dinner	2

Chapter 11

Dental Care



Tooth decay and dental erosion can affect the teeth of any child. Children and adults with Urea Cycle Disorders are at greater risk and need to take good care of their teeth. Sugary foods and acidic drinks may be consumed more often as the diet must be low in protein and high in carbohydrate, which increases the risk of tooth decay. The synthetic protein is acidic and may be sweetened, but remains an **essential part** of your child's diet.

Suggestions for reducing dental problems include:

- Encourage your child to finish their synthetic protein/calorie drink in one go rather than sipping over a long period of time.
- Drinking the synthetic protein/calorie drink (if applicable) with a straw may be beneficial as it reduces exposure of the teeth to sugar and acid.
- It is best to consume synthetic protein/calorie drink at meal times. Give some water after each synthetic protein/calorie drink.
- Water is the best drink to have apart from the synthetic protein/calorie drink. Encourage often throughout the day.
- After the age of 6 months, encourage your baby to start using a beaker. The main reason for this is that bottle feeding can be harmful to teeth.
- Only put infant formula and water in an infant's bottle.
- Do not let your child sleep with a bottle or beaker in their mouth.
- If sweets, juices or fizzy drinks are taken, it is best to take them with meals, rather than between meals.
- Visit your dentist regularly, at least once every year. It might be worth discussing with your dentist preventative measures to reduce decay such as fissure sealants.
- Do not use toothpaste until your child reaches 2 years of age. Use a soft toothbrush and water only once their teeth appear.
- From age 2-7 years use a small pea size amount of fluoride toothpaste.
- We recommend to assist your child with brushing their teeth up to the age of 8 years.
- Ensure regular brushing of teeth twice per day. Brushing teeth should last for 3 minutes.

See: <http://www.dentalhealth.ie> for further information on dental health.

Chapter 12

Teenagers

During the teenage years, many questions may arise as your teenager becomes more independent with their diet and medications. There is a move of responsibility of managing the diet and medications from the parent to the young adult. This chapter looks at some of the issues that may occur during the teenage years.

The Urea Cycle Disorder diet and medications are for life

With Urea Cycle Disorders, there is a risk of serious consequences if your teenager overexchanges or does not take the full amount of their synthetic protein or medications. Taking extra synthetic protein can also have consequences so it is important to stick to the recommended amount. It is important that they understand the effects of not strictly following their diet and treatment. The dietitian and metabolic team will work with your teenager to help fit their diet and treatment into their lifestyle.

Guidelines for teenagers

Take medications as recommended. Understand the role of the different medications they are on and the importance of them.

Take the synthetic protein (if applicable). Make sure they take the full amount every day and spread throughout the day (ideally over 12 hours). If they take energy, vitamin or mineral supplements take these daily as recommended by the metabolic team. These supplements are important to the control of their Urea Cycle Disorder and for their health and wellbeing.

Count protein exchanges as directed by the metabolic team, spread them throughout the day. Don't save protein exchanges up for one meal.

Understand any changes needed when they are **sick**, ensure that they are familiar with their unwell plan and how to use this when they are sick.

Attend the metabolic outpatient appointments so that the team can update their treatment and their diet can be checked to make sure it is adequate.

Even though your teenager has a Urea Cycle Disorder they should aim to follow healthy eating guidelines within the restrictions of a low protein diet.

It is important that your teenager understands about their Urea Cycle Disorder and all parts of its management. You should encourage them to read up about their Urea Cycle Disorder and ask the metabolic team to explain anything they do not understand.

If they have not been to clinic recently they should not be embarrassed about getting back in touch. The team will be delighted to see them again and bring them up to date with the management and treatment of Urea Cycle Disorders.

They need to be aware that Urea Cycle Disorders can cause them to get very sick if they become unwell (for example, catch the flu), have an accident or need an operation. They need to clearly understand the changes needed to their diet and medications when unwell.

Exercise and sport

Having a Urea Cycle Disorder does not limit their ability to participate in exercise or sport. Regular physical activity is an important part of a healthy lifestyle and we encourage it. The Irish recommendations for exercise are moderate activity for at least 60 minutes every day for teenagers. For example; cycling, running, boxing, martial arts, hurling/camogie, brisk walking, football, skate boarding, dancing, skipping, rugby, tennis, swimming and basketball.

After exercise, the body needs fluid, carbohydrate and protein to recover. Drink plenty of fluid, especially water and eat some carbohydrate foods like low protein bread or pasta. The type, intensity and length of time exercising will determine how much protein they require.

Taking their natural protein (exchanges) and synthetic protein (if applicable) as prescribed should provide their body with enough protein. 'Protein powders' that promise to increase muscle bulk are **not appropriate** for a person with a Urea Cycle Disorder as they will increase their blood ammonia levels. Ask the dietitian if additional protein is required.



Eating out and alcohol

These topics will be discussed in chapter 13.

Pregnancy

Pregnancy will affect control of Urea Cycle Disorders. This will require changes in their treatment, blood monitoring and diet during pregnancy and immediately after the birth. For more information see later chapter 15. We would advise all pregnancies for patients with Urea Cycle Disorders to be planned and to talk to the metabolic team in advance.

Chapter 13

Eating Out & Alcohol

Eating Out

Eating out with Urea Cycle Disorders can be enjoyable and easy if you take the time to prepare in advance. Many restaurants have menus online so you can view these in advance. It is a good idea to phone ahead and explain a little about the diet in order to find out if the restaurant has suitable dishes already on their menu. Some menu items may need slight modification such as leave out the cheese topping. Find out if they would be willing to cook low protein pasta, bread or pizza bases.

When eating out there are two options you can take:

1. Stick to your usual pattern of spreading your exchanges throughout the day, meaning you have only a few left for dinner.
2. Save up your exchanges for the meal if it is a special occasion, have all low protein meals during the day. **This should not be done on a regular basis.**

The following are ideas for eating out in different locations:

Barbeques

- Homemade vegetable and fruit skewers
- Vegetable kebabs marinated in garlic and honey sauce or other low protein marinades
- Salads*
- Hash browns*
- Vegetable fingers*
- Vegetable burgers*
- Corn on the cob*
- Mushrooms*
- Aubergine slices
- Dips, e.g. salsa, chutney, guacamole*

**May need to count exchanges*



Cafes

- Mixed salad with a suitable dressing and coleslaw
- Fruit salad/fruit juice
- Chips*
- Jacket potato* with low protein filling – tomato, coleslaw, sweetcorn*, guacamole* and sweet chilli sauce
- Vegetable soup (not based on milk, cream, lentils or other beans, e.g. kidney or white beans or chickpeas)
- Mashed potato* and vegetables with gravy
- Latte, cappuccino, milkshake, smoothies made on coconut milk or rice milk (**Note: Rice milk is not suitable for children under 5 years and pregnant or lactating women**)

Italian restaurants

- Pasta** with tomato based sauces
- Garlic mushrooms* on garlic bread**
- Garlic bread**
- Bruschetta**
- Vegetarian antipasto – artichokes, olives, tomatoes, roasted peppers, tapenade
- Pizza**
- Gluten free pizza base* (check with the restaurant) with low protein cheese**
- Fruit
- Sorbet

Chinese restaurants

- Boiled rice*, plain fried rice*, noodles*
- Vegetable dishes, e.g. stir fried vegetables
- Prawn crackers*
- Vegetable spring rolls*
- Vegetable Chinese soup*

**May need to count exchanges*

***Bring your low protein product for the restaurant to prepare*

Thai/Malaysian restaurants

- Boiled rice*, plain fried rice*, noodles*
- Vegetable dishes, e.g. green or red curry (avoid dishes with nuts and cream based sauces, e.g. panang curry)
- Fresh fruit

Indian restaurants

- Boiled rice*
- Dishes made with sago*
- Vegetable dishes without legumes (lentils, dahl, red or white beans, chickpeas) or paneer (indian cheese)
- Mango chutney and pickle, lime pickle
- Poppadums*

Greek restaurants

- Rice*
- Dips – hummous*, baba ganoush (check ingredients)*
- Olives, cucumber, red onion and tomato in a vinaigrette
- Vegetable dishes, e.g. vegetable kebab
- Salad

Mexican restaurants

- Sweet potato wedges*
- Tortilla chips*
- Corn chips*
- Taco shells*
- Salsa
- Guacamole*
- Stir fried seasoned vegetables

**May need to count exchanges*

Alcohol

Having a Urea Cycle Disorder does not stop you from drinking alcohol. The minimum age alcohol can be legally consumed in Ireland is 18 years. The current guidelines in Ireland recommend a maximum of 17 units of alcohol for men and 11 units for women per week. It is recommended to have at least two alcohol free days each week. Drinks should be spaced out over the week and not consumed in one sitting. Drinking more than the recommended amount may cause harm.



What is a unit?

Small glass of wine (100ml)

1 pub measure of spirit (35.5ml)

½ pint of beer, ale, cider, Guinness

1 alcopop (275ml)

There are many suitable alcoholic drinks for people with Urea Cycle Disorders. However, some alcoholic drinks contain natural protein and therefore should be restricted.

Suitable alcoholic drinks for Urea Cycle Disorders

- Cider-dry, sweet and vintage such as Ritz or Bulmers
- Glass of red/white/rose wine (need to count if taking more than 2 large glasses of wine) *Note: 1 bottle (750ml) of wine is approximately 1 exchange*
- Fortified wine-port and sherry
- Liqueurs-cherry brandy and curacao
- Vermouth-dry and sweet
- Spirits-whiskey, gin, vodka, rum, brandy, pimm's and martini
- Alcopops-Bacardi breezer and WKD
- Cocktails including suitable alcohol



Chapter 14

Overseas Travel

Being on a low protein diet does not mean that holidays should be restricted to locations that are close to home. There is no reason that anyone with a Urea Cycle Disorder, with some forward planning, cannot travel anywhere in the world and have a great holiday.

When you have a Urea Cycle Disorder, travelling requires careful planning to ensure you have your low protein foods, synthetic protein (if applicable), medications and other equipment to hand at all times. **A known metabolic centre should be located within a safe travelling distance of your holiday destination in the event of illness.** The metabolic team can provide you with a list of metabolic centres worldwide and their contact details. You will also need to bring emergency letters in case you become unwell as well as supply of products for the unwell plan and extra medications. The more often you travel the easier this planning becomes and the more confident you become.

Dietetic customs letter

If you are travelling overseas you must take a dietetic customs letter. The customs letter will list the name of the synthetic protein (if applicable), low protein foods and products for the unwell feeds that you will be carrying in your luggage. If you do not have this letter you may have difficulties in taking your necessary products into another country. **Ensure to give the metabolic dietitian plenty of notice to arrange this letter.**

Emergency medical letter

You should also take a copy of your emergency medical letter with you which lists your medications and a copy of the unwell plan. You need to request this from the doctors in the metabolic team.

Travel insurance

Ensure to take out travel insurance.

Vaccinations

Make sure all vaccinations are up to date and you or your child all have the appropriate vaccinations and precautions for the countries you are travelling to.

Packing your suitcase

Ensure you keep the synthetic protein (if applicable) in its original sealed packaging. Some people find it useful to change to a powder version of their synthetic protein to reduce the overall weight of luggage. You may be able to ship your product to your destination in advance. Contact the nutrition company who makes the product for further details on this.

If you are flying, you can contact the airline to enquire about extra baggage that may be required as some airlines allow you to bring it free of charge.



Lost luggage

It is a good idea to carry extra synthetic protein (if applicable) and medications to cover the possibility of your luggage being lost. Divide it between your suitcase and hand luggage. Make sure you have enough synthetic protein, low protein foods and medications to last a few days in case you arrive at your destination before your main luggage.

Arrange to have someone on standby to post any items you may have forgotten.

Eating on the flight

Contact your airline to see if suitable food options are available. Take low protein foods and exchange foods for the flight. Be prepared for flight delays. It is a good idea to have a plan for the first meal when you arrive at your destination. Have your synthetic protein and medications within easy access.

Booking accommodation

When booking your holiday, it is important to look at your accommodation options.

- Hotel accommodation – before travelling, enquire about the type of meals the hotel provides. A buffet style meal is preferred as it offers a wide variety of options. Contact the accommodation in advance and ask them if they will be willing to prepare your low protein foods. It may be a good idea to provide them with some simple recipe ideas.
- Self-catering may be an option. Check the cooking facilities and local supermarkets that are available.

Label reading

Food labels may be different in foreign countries. Take a dictionary to help with translation.

Useful phrases

You may wish to print off some useful phrases and explain Urea Cycle Disorders and dietary requirements in the language of the country you are travelling to. These could be shown in restaurants and hotels.

Travelling for long periods

If travel is planned for long periods, for example, a few months or longer, this will need to be organised at least 6 weeks in advance. You will need to contact the manufacturer of both your low protein foods and synthetic protein to see what services are available. There is no guarantee that your synthetic protein and low protein foods will be sent to your required destination free of charge. This is why forward planning is essential.

If you are planning to stay in one destination for your trip you can be linked in with the nearest metabolic centre, to have your bloods done and arrange any outpatient follow up if required.

Holiday Checklist

1. Dietetic customs letter and emergency medical letter
2. Synthetic protein including some extra in case of emergencies (if applicable)
3. Low protein foods
4. Products for the unwell feeds in the event of illness
5. Medications including extra in case of emergencies
6. Mixing container and scoop measures
7. Dictionary
8. Diet information for example exchange booklet, calculator, recipes
9. Hospital contact number in Dublin
10. Hospital contact number in destination country
11. Passport :)



Chapter 15

Pregnancy

As Urea Cycle Disorders are rare, there are only small numbers of women worldwide who have had their own children. When you are planning a pregnancy, talk to your metabolic team about the issues for you and your pregnancy. Also discuss the risk of your baby having a Urea Cycle Disorder. Some changes and precautions will be necessary. It is important to:

- Be healthy before you get pregnant. Have your Urea Cycle Disorder 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.
- All women planning pregnancy should take a 400µg folic acid supplement for three months prior to conception and for the first 12 weeks of pregnancy.
- You may also require DHA (docosahexanoic acid), a special type of omega 3 fat, before and during your pregnancy.
- Maintain close contact with the metabolic team before and during pregnancy. Let the team know as soon as you know you are pregnant.
- You will need more frequent blood tests-the metabolic team will advise you on frequency of these tests. Changes to your diet and amount of synthetic protein will be necessary as your pregnancy progresses. You will need more natural 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.
- If you are suffering from morning sickness, inform the metabolic team. Your metabolic doctor and dietitian will work together with you to come up with a plan to ensure you get enough natural and synthetic protein and energy to help maintain good blood ammonia and amino acid levels.



- Gain sufficient weight during the pregnancy (discuss this with your dietitian). You will need to use additional low protein foods and may need energy supplements. Adequate energy intake is important during pregnancy to keep good blood ammonia and amino acid levels and also for your baby's growth.
- During the labour and immediately after the birth, it is likely that you will need similar management to that used during illness for two to three days. Changes to the uterus can rapidly increase blood levels of ammonia. Your metabolic team will discuss management of this with your obstetrician.
- Breastfeeding should be possible with regular blood monitoring to check that your intake of protein and energy is adequate.
- Follow the unwell plan during illness and notify your metabolic team if you are unwell.

Obstetric care during the pregnancy

Your obstetric care before the birth is similar to women without Urea Cycle Disorders. You will be in more regular contact with your metabolic team. Your metabolic team will advise your obstetrician, GP or midwife about the treatment for your Urea Cycle Disorder during the pregnancy. They will also liaise closely with the maternity hospital on the plan around the time of the birth.

Like all babies born in the Republic of Ireland, your baby will be tested for many different metabolic disorders and other conditions soon after birth. Additional tests need to be done to check your baby for Urea Cycle Disorder. The metabolic team will advise you and the hospital when the tests should be done and if any special care is needed for your baby.

Contact the maternity hospital where you will be having your baby well before your due date to plan your diet during the admission. The metabolic dietitian can help you plan your meals with the dietitian in the maternity hospital.

Breastfeeding

Breastfeeding is ideal for babies and has many health benefits for both you and the baby. Breastfeeding should be possible. You will most likely need to eat more natural protein and energy than before you were pregnant. Regular blood monitoring will check that your intake of protein and energy is adequate.

If you are planning to breastfeed, please discuss this with the metabolic dietitian.

Chapter 16

Health Related Benefits & Entitlements

A medical social worker will meet with all newly diagnosed Urea Cycle Disorder patients and their families. The Medical Social Worker can be contacted on 01 8784212.



Chapter 17

Glossary

Amino acid

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

Ammonia

Ammonia is a chemical formed in the liver when excess protein is eaten. Normally, the ammonia is changed through a series of steps (the urea cycle) into another chemical called urea. Urea will pass through the kidneys where it is removed from the body in the urine. High levels of ammonia are toxic and it can enter the brain and damage it. If left untreated, high ammonia levels can be life threatening.

Calorie

A calorie is a measure of energy.

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 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.

Essential Amino Acid Supplement

Also called synthetic protein. This can be taken by people with a urea cycle disorder to replace the protein in their diet. It contains all essential amino acids and some contain vitamins, minerals. A *set amount* of this is given.

Exchange

An exchange system is used to count protein in the diet. One exchange = 1 gram of protein. Also known as natural protein. People with Urea Cycle Disorders will be on a set amount of exchanges to provide the body with enough protein for growth and development. This will vary depending on blood results.

Fasting

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

Folic acid

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

Gene

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

Natural Protein

Also known as exchanges. People with Urea Cycle Disorders will be on a set amount of natural protein to provide the body with enough protein for growth and development. This will vary depending on blood results.

Protein

Protein is made up of amino acids. It is needed by the body for growth and repair. Many foods contain protein. Foods such as meat, fish, chicken, eggs, milk, cheese, yogurts, soya, nuts, bread, pasta and chocolate are rich in protein and are not suitable for people with Urea Cycle Disorders. A *set amount* of natural protein and synthetic protein may also be recommended to ensure adequate intake of essential amino acids.

Synthetic protein

This may be given to people with Urea Cycle Disorders to ensure they get enough essential amino acids in their diet. Vitamins and minerals may be included. A *set amount* of this is given.

Urea Cycle Disorders

A group of metabolic disorders involving the urea cycle. This cycle is responsible for clearing the body of ammonia produced from the breakdown of protein.

Acknowledgements:

All members of the National Centre for Inherited Metabolic Disorders (NCIMD) team who contributed to this guide.

ASIEM (Australasian Society for Inborn Errors of Metabolism)









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