



Metabolic.ie

National Centre for Inherited Metabolic Disorders



Homocystinuria

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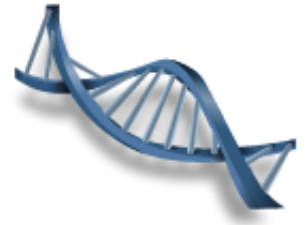
Introduction

The aim of this guide is to provide information for individuals with HCU or for parents/carers with a child who has HCU. 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 HCU...?



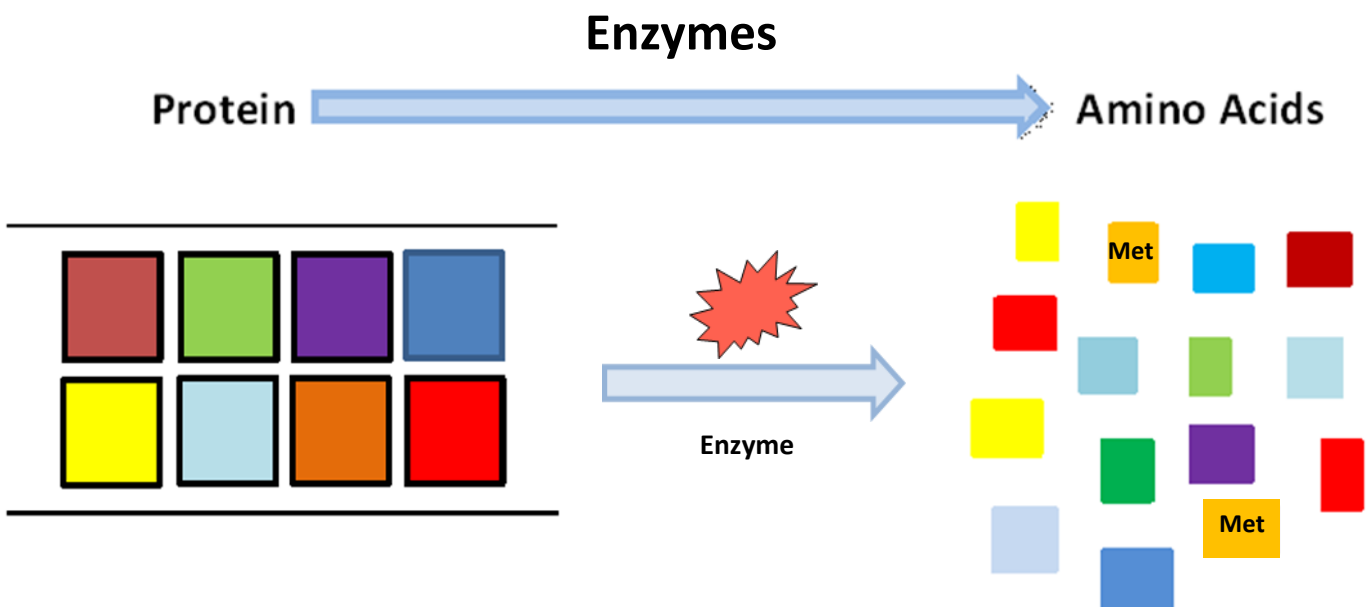
Homocystinuria (HCU) is an inherited metabolic disorder which affects the breakdown of protein containing foods.

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 including growth, repairing cells and tissues as well as making other body proteins.

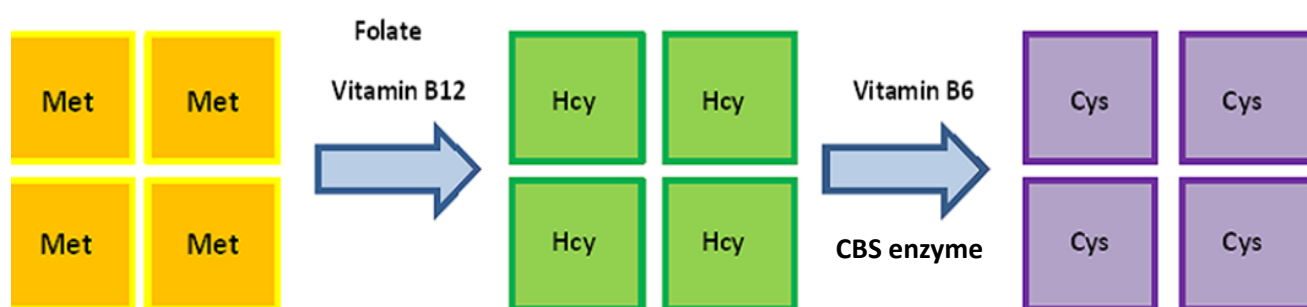
Methionine (Met) is one of these amino acids. People with HCU are born without or with very little of the **enzyme** cystathionine beta synthase (CBS), which is used to change methionine to another amino acid called cysteine. This causes methionine and another substance called homocysteine (Hcy) to be high and the cysteine to be low in the blood.

There are two different forms of HCU. One form can be treated with vitamin B6 supplements (also called pyridoxine) and this type is called 'B6 responsive HCU'. The other form of HCU cannot be treated with vitamin B6 supplements alone and this type is called 'non-B6 responsive HCU'. This booklet will cover the dietary management of '**non-B6 responsive HCU**'.

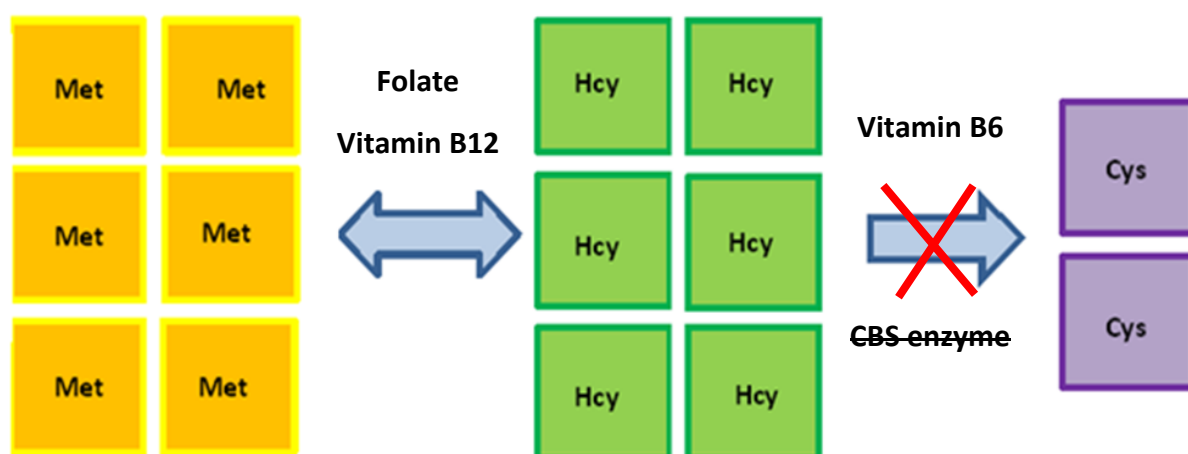
If left untreated over a period of time, high homocysteine levels can cause problems to several parts of the body which include; blood clots, dislocation of the lens of the eye, learning disability and bone problems.



In a person **without** HCU, methionine is converted into homocysteine and then into another amino acid called cysteine. Folate, vitamin B12 and vitamin B6 help this reaction.



In a person **with** HCU, the conversion of methionine into cysteine does not happen as it should, causing methionine and homocysteine levels to rise and cysteine levels to be low.



How is HCU treated?

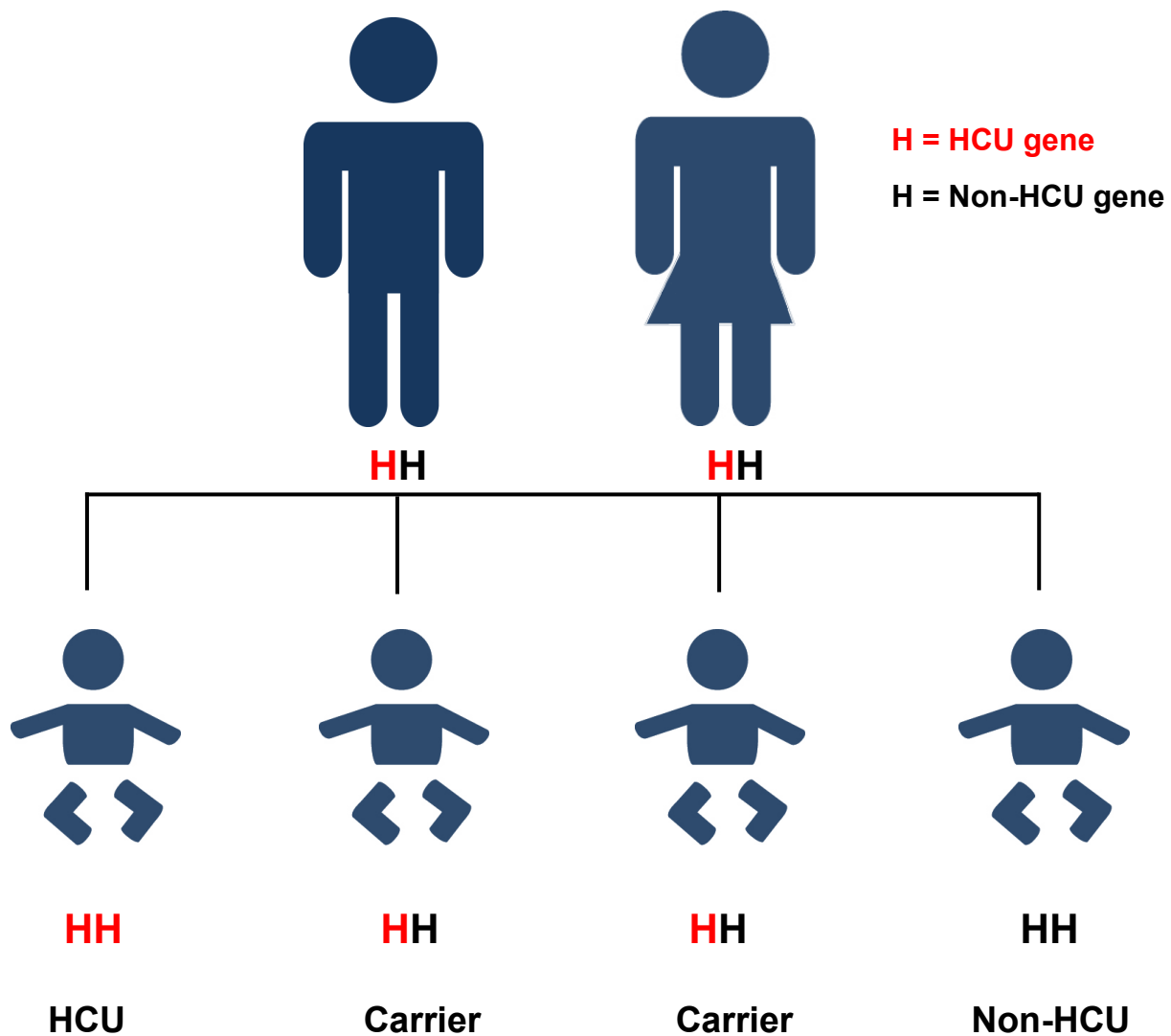
The main aim of treatment is to ensure acceptable levels of methionine, homocysteine and cysteine and to provide adequate protein to support normal growth and development. **The treatment of HCU is a low protein diet for life, in addition to a methionine-free supplement** (synthetic protein). The synthetic protein contains other amino acids including cysteine, vitamins and minerals your child needs. Supplementation of B6, B12, folate, cysteine and betaine may be recommended.

An essential part of the treatment of HCU is to do blood samples regularly to measure homocysteine levels in the blood. Methionine and cysteine levels are also checked during these blood tests. These bloods are sent to the Metabolic Lab in Temple Street. **We aim to keep total homocysteine levels less than 100µmol/L.** Blood levels need to be done regularly depending on your child's age and levels.

HCU is a treatable disorder. When detected and treated early, children with HCU are able to reach their full potential. Provided your child's HCU is well controlled, they have every chance of attending university, building a successful career and enjoying a happy family life just like everyone else.

How is HCU diagnosed and what causes it?

All babies born in Ireland are screened for HCU by the heel prick test shortly after birth. This picks up **most but not all** babies with HCU. Around one in 65,000 babies born in Ireland have HCU. HCU is an inherited metabolic disorder. Children inherit two sets of genes, one from each parent. As a parent of a child with HCU, you have one HCU gene and one non-HCU gene. This is known as being a carrier. People who are carriers for HCU do not have HCU themselves. Your child has inherited two HCU genes, one from their mother and one from their father. This means that your child has HCU. **It is important to remember HCU 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 HCU.



Chapter 2

Overview of Treatment



The National Centre for Inherited Metabolic Disorders (NCIMD) is where all children diagnosed with HCU are treated in the Republic of Ireland. At present, HCU is treated by diet and medications. HCU should not restrict your child or your family from any normal activities, for example going on holidays, eating out or playing sport.

Successful treatment of B6 non-responsive HCU 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 methionine and are not allowed in the diet.

Natural protein/Exchanges: People with HCU still require a small amount of methionine 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.

Daily intake of synthetic protein: This contains amino acids (except methionine) and vitamins and minerals. It can be given in the form of a formula, drink or gel. The synthetic protein should be taken several times throughout the day. The amount required will be calculated specifically for your child's needs.

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.

Medications: Pyridoxine (vitamin B6), vitamin B12 and folic acid are medications that help reduce the amount of homocysteine in the blood and are often used as part of the treatment of HCU. Betaine and cysteine may also be recommended by your metabolic doctor.

Blood levels: Regular monitoring of your child's blood homocysteine, methionine and cysteine is an essential part of the overall management of HCU. Your metabolic team will let you know how often blood levels need to be done. **We aim to keep total homocysteine levels less than 100µmol/l.** Depending on your child's age, frequency of bloods can vary from weekly when your child is younger, to monthly as they get older.

Keeping supplies up-to-date: It is very important to keep an up-to-date supply of the synthetic protein, low protein foods, medications and blood 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 HCU will have high homocysteine and methionine levels initially. First, your baby was given a trial of vitamin B6 (pyridoxine) supplements for a number of days. As your child was non-B6 responsive, the B6 did not improve levels and therefore your child needs to start a special diet. To reduce homocysteine and methionine levels, your baby will be given a methionine-free formula (HCU infant formula) instead of breastfeeds or standard infant formula for the first few days after diagnosis of non-B6 responsive HCU. This will allow the homocysteine and methionine blood levels 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 two different formulas. These should always be given separately and not mixed together.

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 blood homocysteine and methionine at an acceptable level. Breast milk contains less methionine than standard infant formula. However, breast milk **alone** contains too much methionine for babies with HCU. Therefore, HCU infant 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 HCU infant formula to give. After the HCU infant formula, 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. A measured amount of HCU infant formula is given by bottle first to reduce the amount of methionine taken from breast milk.

A measured amount of HCU infant formula is 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 HCU infant formula.

While breastfeeding, you do not need to alter your diet in any way because your baby has HCU. 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 of non-B6 responsive HCU 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 to 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 HCU infant formula. A measured amount of standard infant formula is given first and then HCU infant formula to appetite.

A measured amount of standard infant formula is given first and then HCU infant formula to appetite.

If your baby is hungry in between feeds you may give extra HCU infant formula in addition to the measured amounts of standard infant formula.

HCU infant formula (synthetic protein)

HCU infant formula contains all the nutrients needed for growth, except methionine. This will be prescribed for your baby.

The amount of HCU infant formula 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 will advise you on this as your baby grows.

Preparing formula (standard infant formula or HCU infant 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

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

To store the formula

- Make up each bottle as required. 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 HCU infant formula 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	HCU infant formula (measured)	Breastfeed (to appetite)
7am	Feed 1	30ml	Right side
10.30am	Feed 2	30ml	Left side
1.30pm	Feed 3	30ml	Right side
2.30pm			Left side
4pm	Feed 4	30ml	Right side
7pm	Feed 5	30ml	Left side
10.30pm	Feed 6	30ml	Right side
1.00am	Feed 7	30ml	Left side
4am	Feed 8	30ml	Right side
5am			Left side

For formula-fed babies:

Feeds

Date.....

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

For breastfed babies:

Feeds

Date.....

[illegible]

For formula-fed babies:

Feeds

Date.....

[illegible]

Chapter 4

Understanding the Synthetic Protein

Synthetic protein is a specially made protein that contains all of the amino acids (except methionine) that your child needs to grow, build and repair tissues. The synthetic protein will also contain all of the vitamins and minerals that your child needs for good health.

The synthetic protein is an essential part of the treatment of HCU. If it is not taken as recommended, homocysteine and methionine levels will rise, 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, homocysteine and methionine levels.

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

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 available 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.

The synthetic protein must be spread throughout the day. This helps keep homocysteine and methionine levels within an acceptable range. If your child is struggling to take all of their synthetic protein, please inform your dietitian. **As HCU requires a diet for life, the synthetic protein must also be taken for life.**

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 HCU is a low protein diet. **This is a diet for life.** The diet prevents a build-up of excess methionine and homocysteine in your child's blood, while at the same time providing enough protein for normal growth and development. There are four main components of the diet:

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

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

RED STOP:

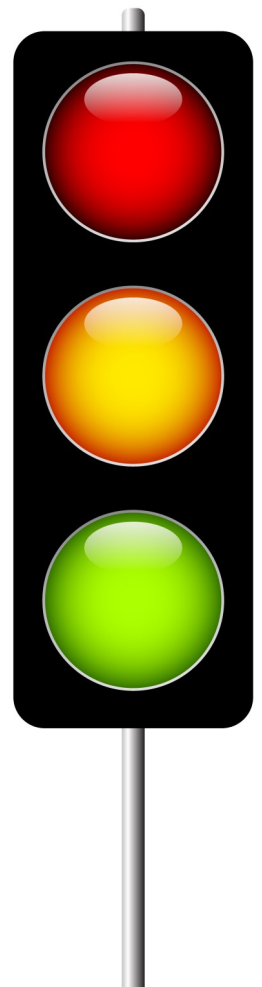
Do not eat these foods as they are too high in natural protein (methionine).

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.

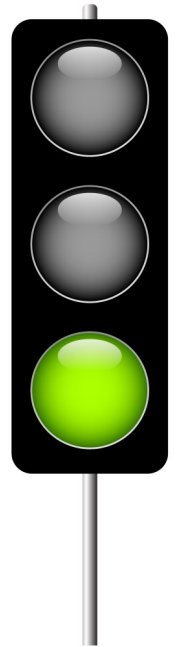
There is a large variety of specially manufactured low protein foods such as bread, pizza bases, pasta, rice, flour, cereals, milk, meat substitutes, cake mixes, biscuits and chocolate. These foods are available on prescription from your local pharmacy.

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 HCU are encouraged not to have too much sugar and fat for the same reasons as other children without HCU.

The following tables give examples of foods that are naturally low in methionine 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
White	Boiled sweets	Chutneys
Brown	Candyfloss	Golden syrup
Castor	Lollipops	Jam-any type
Glucose	Sorbet	Marmalade
Icing sugar	Sherbet	Treacle
Jam sugar	Iced lollies	Honey
Barley sugar	Suitable jellies*	Maple syrup
	Any chewy sweets*	
	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 konjac (Asian root vegetable)	
Salt	Sago	
Spices	Tapioca	
Vinegar		

Fats and oils	Drinks	Sauces
Butter	Some coconut milks	Apple sauce
Lard	Pure fruit juice	Balsamic vinegar
Margarine	Hemp milk	Cranberry sauce
Suitable low fat spread	High energy drinks	Chutney
Coconut oil	Minerals	Horseradish sauce
Duck Fat	Rice milk (not suitable in children under 5 years or pregnant/ lactating women)	Ketchup, brown sauce
Goose fat	Squashes/cordial	Most mayonnaises, salad creams
Fry light spray	Tea/coffee/herbal tea	Mustard
Grape seed oil	Water/ soda water/mineral water	Pasta sauces, cook- in sauces
Garlic oil		Piri Piri sauce
Olive oil		Relish
Rapeseed oil		Salad dressing
Sunflower oil	See chapter 13 for suitable alcoholic drinks	Soya sauce (check label)
Sesame oil		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 methionine for growth, development and repair. Foods such as cereals, rice, beans, peas and potatoes contain a small amount of protein and methionine. 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 homocysteine 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.

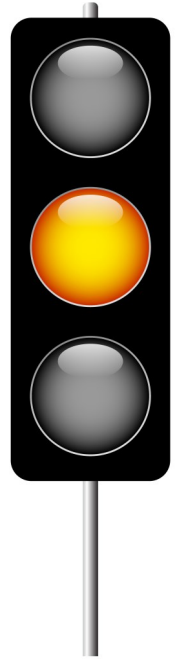
An exchange is the amount of any food that contains 1 gram of protein.

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 methionine. 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

These foods are high in protein and methionine 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.

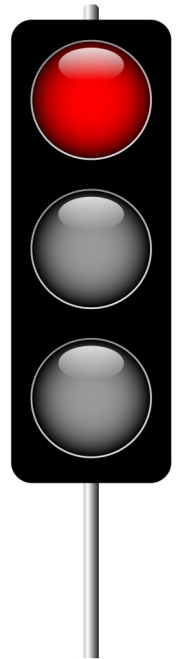
Nuts and seeds: Including peanut butter and nut spreads.

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.

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



Eating the wrong foods will not make your child immediately sick but will cause their homocysteine levels to go high.

Giving unsuitable foods can also result in the child developing a taste for these foods and make the diet more difficult for the child to stick to.

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 on 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 (½carton)
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 homocysteine levels is an important part of managing HCU. Keeping blood homocysteine levels under control reduces the risk of problems. The ideal blood level for total homocysteine is less than 100 μ mol/l.

HCU requires frequent bloods to monitor the effectiveness of treatment. The results of these bloods 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, prior to surgery, air travel and pregnancy. The metabolic team will decide on this and inform you what is necessary at a given time.

Blood samples will need to be taken in your local hospital. The sample needs to be handled in a prompt and appropriate manner to guarantee accurate results. **Bloods need to be treated in a laboratory within 15 minutes of being taken.** The laboratory in Temple Street Children's University hospital has a detailed protocol for this procedure which they will provide to external hospitals if requested. If your child is having their bloods taken in an external hospital they should be informed of this protocol. These bloods will then be sent to the Metabolic Laboratory in Temple Street for analysis. The metabolic nurses will give you the necessary request forms for these bloods at your clinic visits.

If homocysteine levels are too high or too low, your metabolic dietitian will advise you appropriately on changes to make. A number of factors cause blood homocysteine levels to increase or decrease outside of the target range.

Homocysteine levels may increase when:

- Methionine intake (exchanges) is too high.
- Intake of synthetic protein is too low which causes the body to break down its own muscle tissue for nutrients. This releases methionine and homocysteine into the blood.
- A child's rate of growth has slowed meaning less methionine (exchanges) is being used to build new proteins.
- Illness, vaccinations and teething may make it difficult to eat or take the synthetic protein. This can cause higher methionine and homocysteine levels from the breakdown of the body's own muscle tissue.

Homocysteine 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 methionine and homocysteine into the blood.

Homocysteine levels may decrease when:

- Intake of methionine (exchanges) is lower than recommended.
- Growth in a child causes levels to fall as methionine is used to build new body tissues.
- Sometimes following illness when the body is rebuilding body protein.

It is really important to inform your dietitian of any issues that may have arisen in your child during the days prior to levels being taken e.g. over exchanging, illness etc.

Frequency of monitoring blood levels

How often your child's blood is monitored depends on their age and blood levels. This is determined by the metabolic team.

Ringling for levels

It is really important to call the metabolic dietitians for your child's results. The contact information for the dietitians is as follows:

01-8784317 and press 2 for dietitians

If the phone is not answered, please leave a message and we will return your call as soon as possible. The voicemails are checked daily Monday to Friday. To provide an efficient service please ring between the following times:

Monday, Tuesday, Thursday and Friday: 2-4pm
Wednesday and Thursday: 10am-12.30pm



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

Chapter 7

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 HCU diet**' before the time comes to wean and will discuss weaning in detail with you. Ask your dietitian for this booklet.

This booklet can also be downloaded from:

<http://metabolic.ie/patient-family-information/metabolic-conditions/homocystinuria-hcu/>



Chapter 8

Preparing for Childcare, Pre-School & School

Preplanning

Children with HCU 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 HCU with the childcare or school.

Discussing HCU 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 and why they require medications.

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 HCU and the importance of the low protein diet and medications.

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.
- Staying on a protein restricted diet keeps the child healthy and supports development.
- 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.

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 in school

Your child is likely to be happy to take the synthetic protein 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 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
- 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 9

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 HCU 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 he or she grows.

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 HCU has a number of advantages. It:

- Encourages **acceptance**.
- Helps them develop a good **understanding** of HCU.
- 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 HCU 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 HCU. 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
- Know they need to take medications
- Know they need to have blood tests



Toddlers: 2–5 years continued

- Know to check new foods with their parents
- Watch you prepare the synthetic protein
- Start to learn about yes/no foods
- Are aware of exchange foods that need to be counted

Early childhood: 5–8 years

- Understand that they have a condition called HCU
- Understand that they cannot eat high protein foods
- Have a basic knowledge of why the synthetic protein is important
- Know that the medications are important
- Know that they need to go to hospital regularly to have their blood tests done
- Know what is considered a high homocysteine level
- Help you prepare the synthetic protein
- 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



Late childhood: 8–12 years

- Have a basic understanding of HCU, homocysteine, methionine, cysteine and their diet
- Are able to prepare their own synthetic protein
- Understand that taking synthetic protein helps provide nutrition and helps ensure good blood levels
- Understand that taking their medications 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 the recommended range for homocysteine levels

Early teens: 12–14 years

- Have an understanding of what is HCU, homocysteine, cysteine and amino acids
- Understand the risks and effects of high homocysteine levels on their health
- Prepare their own synthetic protein all the time
- Prepare medications with assistance
- Know the recommended range and frequency for blood homocysteine levels
- 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
- Are able to engage in clinic appointments
- For girls: Are aware of pregnancy risks associated with maternal HCU

Late teens: 14–18 years

- Have a thorough understanding of HCU and its effect on their body
- Prepare their own synthetic protein
- Can measure exchanges accurately
- Are able to make and prepare appropriate meal choices
- Independently remember to go to their own local hospital to get their blood sample done
- Are able to make phonecalls with parents to the metabolic dietitians for 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
- Know how often clinic visits are needed and take responsibility for making their own appointments
- Are able to engage in clinic appointments
- Know how to contact their metabolic team when required
- For girls: Are aware of pregnancy risks associated with maternal HCU



Talking about HCU with your child

Talking to your child about HCU 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 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 HCU, 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.
Are you vegetarian?	I do not mind not eating meat – I have never had it and do not miss it. 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 HCU, you have to be born with it.
How can you drink your synthetic protein?	I have taken a synthetic protein since I was a baby, so I'm used to it. I am used to having synthetic protein. It is like medicine – I need it to be healthy.
Why do you have to take those medicines?	The medicines 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

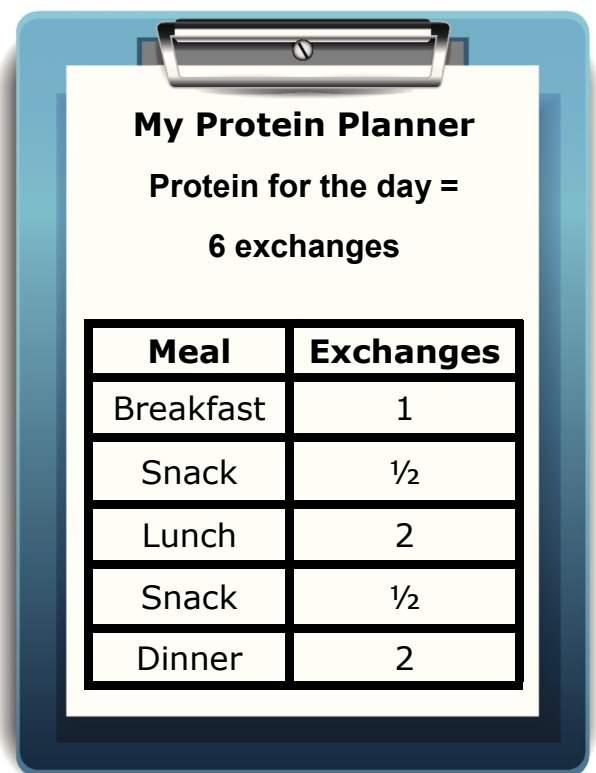
Regular discussions about HCU with your child will help them feel more comfortable talking about HCU and give them the confidence to ask you questions and share their concerns. You can help your child to understand HCU in a variety of ways from an early age. Many families use games, such as the following,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

- Placing different foods into 'yes' and 'no' categories.
- Cutting out different foods from magazines and pasting them into a HCU 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.
- A star chart/reward system for drinking their synthetic protein, managing their diet, taking their medications and doing their blood tests.

The more HCU 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 10

Dental Care



Tooth decay and dental erosion can affect the teeth of any child. Children and adults with HCU 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 in one go rather than sipping over a long period of time.
- Drinking the synthetic protein with a straw may be beneficial (if applicable) as it reduces exposure of the teeth to sugar and acid.
- It is best to consume synthetic protein at meal times. Give some water after each synthetic protein.
- Water is the best drink to have apart from the synthetic protein. 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 11

Illness

Children with HCU will have the same number of coughs, colds and other illnesses as a child without HCU. Illness may cause a temporary rise in your child's homocysteine level, **but no long-term harm is done**. During illness the body starts to break down its own tissues, releasing methionine into the bloodstream, which can cause homocysteine levels to rise. If your child is due to do bloods but is unwell, please contact the metabolic team for advice.

When your baby/child is sick

Contact your GP if your baby/child is unwell just as you would if they did not have HCU. Inform the GP that they have HCU.

Maintaining your child's fluid intake is important during illness. Here are some practical guidelines:

Babies

- Offer feeds more frequently than usual.
- If your baby has a poor appetite and is feeding poorly from the breast you may have to express to keep up your supply.
- Give any medications recommended by your doctor.
- Encourage adequate fluid intake by offering drinks every one to two hours. Fewer wet nappies or strong coloured urine is a sign your baby may not be drinking enough.
- If your baby has gastroenteritis, see your GP as it is important to avoid dehydration.

Older children and adults

- Encourage small frequent meals and regular fluids as tolerated.
- Your metabolic team may advise you to cut back on protein exchanges if homocysteine levels are raised. In many cases this will have happened naturally as appetite may be reduced during illness.
- Ensure adequate fluids are taken especially if symptoms include vomiting and/or diarrhoea. It may be useful to use Dioralyte or an equivalent electrolyte replacement powder to help prevent or correct dehydration.

Older children and adults continued

- Try to ensure the synthetic protein is taken. However do not force it especially if your child is vomiting. If forced, this may lead to problems at a later stage.
- It can be useful to try to dilute the synthetic protein further with water.
- It is also **very important** not to mix any medications such as antibiotics through the synthetic protein as this will alter the taste and may lead to problems at a later stage.

If your child is admitted to a local hospital, inform the metabolic team so that they can liaise with the relevant staff. Remember to bring your child's synthetic protein, low protein foods and medications as they will not be stocked in the local hospital.

If your child is not tolerating feeds for a number of days or requires a surgical procedure, they may need to be admitted to hospital for intravenous (IV) fluids. Please contact the metabolic team for advice.

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 HCU diet and medications are for life

For HCU, there is a risk of serious consequences if your teenager overexchanges or does not take the full amount of their synthetic protein or medications. 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 the synthetic protein, make sure they take the full amount every day and have it three to four times a day with meals so it is 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 HCU and for their health and wellbeing.

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

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

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 HCU 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 HCU and all parts of its management. You should encourage them to read up about HCU 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 HCU.

Exercise and sport

Having HCU 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 synthetic protein as prescribed should provide their body with enough protein. 'Protein powders' that promise to increase muscle bulk are **not appropriate** for a person with HCU as they will increase their blood methionine and homocysteine levels. Ask the dietitian about suitable synthetic protein alternatives and if it is required.



Eating out and alcohol

These topics will be discussed in chapter 13.

Pregnancy

Pregnancy increases the risk of clots in all women. High homocysteine levels also increases the risk of clots. To prevent clotting during pregnancy, women with HCU should ideally plan their pregnancy and aim for good homocysteine levels before they conceive. Pregnant women will require changes in their treatment, blood monitoring and diet during pregnancy and immediately after the birth. Additional support from the metabolic team will be provided during this time. The haematology team will advise on anti-coagulant therapy during the pregnancy and for a number of weeks post-delivery of the baby to protect the mother from having a clot. See chapter 15 for more information.

Chapter 13

Eating Out & Alcohol

Eating Out

Eating out with HCU 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 chick peas)
- 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 HCU 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 HCU. However, some alcoholic drinks contain natural protein and therefore should be restricted.

Suitable alcoholic drinks for HCU

- Cider-dry, sweet and vintage for example Ritz, 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 HCU with some forward planning cannot travel anywhere in the world and have a great holiday.

When you have HCU, travelling requires careful planning to ensure you have your low protein foods, synthetic protein, medications and other equipment to hand at all times. The more often you travel the easier this planning becomes and the more confident you become.

Safety while flying with HCU

Long haul flights are associated with developing clots particularly in your legs. We advise people with HCU to check their homocysteine levels prior to travelling and take the following precautions to reduce the risk of developing a clot:

- During the flight wear compression stockings (Grade II Flight Stockings-available in local pharmacies)
- Move your legs frequently and walk around the cabin when possible
- Drink plenty of fluids before and during the flight
- Avoid alcohol the day before and during the flight
- Avoid sedatives (especially sleeping tablets)

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 and low protein foods 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.**

Medical letter

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

Travel insurance

Ensure to take out travel insurance.

Packing your suitcase

Ensure you keep the synthetic protein 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 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 HCU 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, if there is one, to have your bloods done and arrange any outpatient follow up if required.

Holiday Checklist

1. Dietetic and medical customs letter
2. Synthetic protein including some extra in case of emergencies
3. Low protein foods
4. Medications including some extra in case of emergencies
5. Mixing container and scoop measures
6. Dictionary
7. Diet information for example exchange booklet, calculator, recipes
8. Hospital contact number in Dublin
9. Passport :)



Chapter 15

Pregnancy

Women with HCU have to take particular care before and during pregnancy. This is because both pregnancy and high homocysteine levels increase the risk of clots for the mother.

We recommend that all women with HCU should ideally plan their pregnancies. We also recommend that women with HCU who are planning a pregnancy or are pregnant should maintain blood total homocysteine levels of **less than 100µmol/l**.

If you are thinking about planning a pregnancy, talk to the metabolic team about the issues for you and your pregnancy. Also discuss the risk of your baby having HCU. Some changes and precautions will be necessary. You should start sending more regular blood levels. The metabolic dietitians will advise you on ways to get homocysteine levels in the acceptable range. Once pregnant, you will be referred to the haematology team for advice on anti-coagulant therapy.

We recommend that **all women with HCU should ideally plan their pregnancies**. However if the pregnancy is unplanned, contact your metabolic team immediately.

It is important to:

- Be healthy before you get pregnant. Have your HCU 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. Check with the metabolic team that the folic acid you are on is sufficient.
- 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. Aim to do regular levels prior to conception. Let the team know as soon as you know you are pregnant.
- You will need more frequent blood 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. You may also need to start taking a cysteine supplement.
- You will be referred to the haematology team as soon as you are pregnant. You will need to start on anti-clotting medications (e.g. low molecular weight heparin) during the pregnancy and will need to continue this for a number of weeks after the birth. The haematology team will advise you on this.
- The team will work with your obstetrician (pregnancy doctor) and haematology team 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 synthetic protein, energy and fluids to help maintain good blood homocysteine 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 homocysteine levels and also for your baby's growth.
- If breastfeeding regular blood monitoring will continue in the first few weeks to check that your intake of protein and energy is adequate.



Obstetric care during the pregnancy

Your obstetric care before the birth is similar to women without HCU. The difference is you will be in regular contact with your haematology team and metabolic team. Your metabolic team will advise your obstetrician, GP or midwife about the treatment for your HCU during the pregnancy. They will also liaise closely with the maternity hospital on the plan around the time of the birth. For a number of weeks after the birth, you will need to do more frequent blood levels and liaise closely with both the haematology team and metabolic team.

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. The metabolic team will advise you and the hospital when the tests should be done.

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. 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 your metabolic dietitian.

Chapter 16

Health Related Benefits & Entitlements

A medical social worker will meet with all newly diagnosed HCU 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.

Calorie

A calorie is a measure of energy released when a food is eaten.

Energy

Energy is the capacity of the body to do work. The body obtains its energy from the carbohydrate, fat and protein in food.

Enzyme

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

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 HCU will be on a set amount of exchanges to provide the body with enough methionine for growth and development. This will vary depending on blood results.

Folic acid

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

Gene

Genes carry hereditary information for bodily processes and traits, such as blood group and hair colour, and instructions for producing chemicals.

Homocystinuria

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

Methionine free formula

Also called the synthetic protein. This is taken by people with HCU to replace the protein in their diet. It contains all essential amino acids (except methionine), plus vitamins, minerals and extra cysteine. It is an essential part of the treatment for HCU.

Natural protein

Also known as exchanges. People with HCU will be on a set amount of natural protein to provide the body with enough methionine 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 HCU. A synthetic protein supplement makes up for the protein they are unable to eat in food.

Pyridoxine

Also called vitamin B6, this acts as a helper for some enzymes and may be used in high doses in the treatment of homocystinuria if people are responsive.

Synthetic Protein

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

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ASIEM (Australasian Society for Inborn Errors of Metabolism)





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