

Family Guide for Homocystinuria (HCU)

A brief guide for families dealing with Homocystinuria



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What is HCU?

It is a rare genetic disease that is common in Qatar. This disease affects protein digestion.

The name Homocystinuria refers to having high levels of a substance called homocysteine in the urine and blood.

What happens in HCU?

HCU happens due to the absence of a specific enzyme in the body, where the body is not able to digest all the protein.

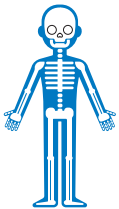
After eating a high protein meal that exceeds requirements, the amino acid Methionine and a harmful substance called Homocysteine levels increase in the blood.

What Happens if HCU is Untreated for a long time?



- **Eyes**

Severe short-sightedness and the eye lens could move out of its normal position.



- **Skeleton**

Tall stature with long legs and arms, curved foot and spine, and increased risk for weakened or broken bones



- **Brain**

Delayed development, decreased mental abilities, seizures, behavioral and psychiatric problems (e.g. anxiety and depression)



- **Blood vessels**







Blood clots (e.g. stroke)

How to monitor blood values?



Homocystinuria is monitored by taking a blood sample to make sure levels of homocysteine and methionine are under control in the blood. The blood sample is taken weekly, once every 2 weeks, once a month or every 3 months depending the age and the blood levels.

Acceptable values:

1- Homocysteine

 Children Below 2 years of age	 Children Above 2 years of age	 Adult
		
Below 20 umol/L	Below 50 umol/L	Below 100 umol/L

2- Methionine

 All ages	Do not forget or miss appointments, regular checkups are very important for the best health outcome and to prevent health problems!
	
18 - 45 umol/L	

How is HCU Managed?

1) Medications used to control HCU

The doctor will prescribe medications with specific doses that can help control homocysteine levels. These medications include:

1. Betaine
2. Vitamins: Folic Acid, Vitamin B12, and Vitamin B6

These medications along with a low protein homocystinuria diet will be used as a complementary treatment to help control homocystinuria.

2) Low Protein Homocystinuria Diet

What is a Low Protein Diet?

People with Homocystinuria should follow a lifelong special diet and it should be low in Natural protein.

Natural protein is the protein that is found normally in most food.

People with Homocystinuria should stop eating foods that are high in natural protein like: meat, chicken, fish, eggs, milk, and milk products. At the same time, they still need a limited amount of natural protein in grams as calculated by the Dietitian that can be taken from other food sources.

For specific protein content of food and their quantities refer to the booklets given by the Dietitian.

Protein Substitute and Medical Food Products for HCU

People with Homocystinuria consume low protein diet, which will not cover the full protein requirement. Therefore, they need additional protein because it is important for the human body functions and growth.

Protein Substitute Formula

The Dietitian will prescribe protein substitutes that are free of the unwanted amino acid (Methionine-free). They are important because they provide the body with protein and energy needed for normal body functions, and they help maintain normal Homocysteine and Methionine blood levels.



Medical Food Products

- Low Protein Medical food will also be prescribed to substitute some of the normal foods that are high in natural protein. Some of these medical foods include rice, flour, pasta, milk substitute and egg replacer/substitute. These products will ensure that growth requirements are met.



Your baby Feeding Journey

Until 6 months of age

It is important to follow the dietitian plan on how to prepare the feed.

Whether the baby is breast feeding and/or infant formula, you need to mix the Protein Substitute milk with a specified amount of the breast milk and/or infant formula.

By this way you won't exceed the amount of protein that would harm the baby.



Continuous follow ups with the dietitian are required to adjust the quantities for feed preparation according to the age, weight, and protein needs.

Weaning (Above 6 months)

At this stage the baby can start eating solid food gradually and slowly based on the baby's acceptance.

Some of the weaning foods containing protein need to be counted in grams, and other food can be given freely (that does not contain protein).

The dietitian will help you understand how much protein the baby needs and how to count it

How can I start feeding food for my baby?

Step 1

Starting from 6 months, give pureed lump-free vegetables and fruits.



You can start with 1-2 teaspoons a day between milk feeds and increase gradually based on the baby's acceptance

Step 2

From 7-9 months, you can provide thicker food in meals.



You can give the baby low protein cereal and rice with vegetables.

Step 3

At 12 months, you can start giving chopped harder food.



(Like chopped fruits and vegetables, low protein pasta with vegetables, and low protein biscuits)

Step 4

After the age of 1 year,



Shifting from bottle feeds to a sipper cup is important.

Sippy cups may encourage the child to drink the protein substitute needed for the growth and development.

The Dietitian will guide you for further information and support.

Your HCU Diet Should Contain:

Food Group	Food Sources	Comments
Very high natural protein (meat, chicken, fish, eggs, milk, milk products)		This food group is prohibited, as it is very high in natural protein.
Moderate Natural Protein (grains, cereals, breads, cakes, legumes, nuts, chocolate, biscuits, chips, crackers, high protein vegetables and fruits)		This group is best limited strictly, and inclusion in the diet, requires discussion with the Dietitian.
Very Low Natural protein (except some fruits and vegetables)		This group includes all food items that contain low to no natural protein at all. *Follow the Dietitian instruction for specific amounts.
Natural Protein Free Food (butter, jam, honey, fruit juices, ketchup, ranch, oils, jelly, candies)		This group includes all food items that contain no natural protein at all. They can be consumed freely too!
Medical Food (low protein; rice, pasta, macaroni, flour, egg replacer, milk)		This group replaces the regular: rice, pasta, flour, milk, to provide energy needs, and to provide enough variety. This group can be consumed freely!
Protein Substitute		This group will help meet protein requirements according to the age group. The specific amount will be prescribed by the Dietitian.

General tips:

1. Protein Substitute Formula can be mixed with any flavor; you can mix with fruit juices, fruits, honey, sugar, rose water, Medical flavored sachet etc.



2. Low protein flour, egg replacers and milk substitutes, all can be used to make our regular dishes, but in a modified low protein version.

A. Low protein flour and egg replacer can be used to bake cakes and bread.



B. Low protein milk can be used to make Karak or cappuccino.



How Can you Design a 5 gram Natural Protein Meal Plan (Sample)

Meal	Menu example	Protein (in grams)
Breakfast	<ul style="list-style-type: none"> Half a cup of normal cornflakes 1 cup low protein milk Half a Banana 	0.5 0 0.5
Snack 1	<ul style="list-style-type: none"> 1 small pack of juice 2 vanilla wafers 	0 0.5
Lunch	<ul style="list-style-type: none"> 1.5 cup vegetable machbous (low protein rice) Vegetable Saloona 	0 0.5
Snack 2	<ul style="list-style-type: none"> A small pack of Chips (read the label to make sure you count grams of protein) 	1
Dinner	<ul style="list-style-type: none"> 1 cup vegetable macaroni (low protein pasta) 3 stuffed grape leaves (low protein rice) 	0.5
Snack 3	<ul style="list-style-type: none"> 1 cups Popcorn 1 cup low protein milk 	1 0
Total		5 grams of protein

Do not forget to take the protein juice or milk as prescribed by the Dietitian!
Ask the dietitian for the specific daily natural protein amount that you need!

How to Read Nutrition Labels

Step 1



Check out the weight of the product on the package.

(It is usually in grams and found either at the back or the front of the package)

Step 2

حقائق تغذوية	
Nutrition Facts	
Informations Nutritionnelles	
12.5 serving per tin	١٢.٥ حصة لكل عبوة
12.5 servir par boîte	حجم الحصة ٤٠ غرام
Serving Size 40g	حجم الحصة ٤٠ غرام
Portion 40 g	
Amount per serving	
Quantité par portion	المصغرات الحرارية
Calories 150	150
نسبة الإحتياج اليومي % Daily Value*	
Total Fat الدهون الكلية 3.2 g / ل	5%
Matières grasses totales	
Saturated Fat دهون مشبعة 0.6 g / ل	3%
Acides gras saturés	
Trans Fat دهون متشعبة 0 g / ل	0%
Acides gras trans	
Cholesterol كوليسترول 0 mg / ل	0%
Sodium صوديوم 2.8 mg / ل	0%
Total Carbohydrate الكربوهيدرات الكلية 27 g / ل	10%
Glucides totaux	
Dietary Fiber الألياف الغذائية 3.6 g / ل	13%
Fibres alimentaires	
Total Sugars سكريات كلية 0.5 g / ل	0%
Sucres totaux	
Added Sugars سكر مضاف 0 g / ل	0%
Protein بروتين 4.4 g / ل	9%
Protéines	
Magnesium مغنسيوم 44 mg / ل	14%
Magnésium	
Zinc زنك 1.3 mg / ل	9%
Iron حديد 1.5 mg / ل	7%
Iron	

Look at the nutrition facts table and notice what is the serving size.

(Written at the top of the table)

Step 3

حقائق تغذوية	
Nutrition Facts	
Informations Nutritionnelles	
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نسبة الإحتياج اليومي % Daily Value*	
Total Fat الدهون الكلية 3.2 g / ل	5%
Matières grasses totales	
Saturated Fat دهون مشبعة 0.6 g / ل	3%
Acides gras saturés	
Trans Fat دهون متشعبة 0 g / ل	0%
Acides gras trans	
Cholesterol كوليسترول 0 mg / ل	0%
Sodium صوديوم 2.8 mg / ل	0%
Total Carbohydrate الكربوهيدرات الكلية 27 g / ل	10%
Glucides totaux	
Dietary Fiber الألياف الغذائية 3.6 g / ل	13%
Fibres alimentaires	
Total Sugars سكريات كلية 0.5 g / ل	0%
Sucres totaux	
Added Sugars سكر مضاف 0 g / ل	0%
Protein بروتين 4.4 g / ل	9%
Protéines	
Magnesium مغنسيوم 44 mg / ل	14%
Magnésium	
Zinc زنك 1.3 mg / ل	9%
Iron حديد 1.5 mg / ل	7%
Iron	

Look at the nutrition facts table and notice how many grams of protein are available in the specified serving size.

Step 4

To determine the protein content in a specific weight. Divide this weight by the serving size and multiply it by the amount of protein in one serving.

$$\frac{\text{Desired weight (grams)} \times \text{Protein (grams)}}{\text{Serving Size (grams)}}$$

Example: Calculate the amount of protein in 10g of oats

$$\frac{10 \text{ g} \times 4.4 \text{ g}}{40 \text{ g}} = 1.1 \text{ g of protein in 10g of oats.}$$

What Can I Eat Outside Home (at Restaurants/Takeaway /Gatherings)?

Nothing can stop you from enjoying time with friends or family!

You will enjoy many dishes if you choose what is suitable for the best health and avoid dishes that harm.

Check these ideas for eating out in different occasions:

• When having a Barbeque choose:

- Vegetable and fruit skewers
- Vegetable kebab
- Salads
- Grilled Corn
- Mushrooms
- Eggplant slices
- Dips, e.g. Baba Ghanouj.
- Juices
- Small amount of chips or grilled potato



• When eating out in a restaurant or café:

- Mixed salad with a non-milk-based sauce
- Chips or Grilled Potato with grilled vegetables
- Vegetable soup (not based on milk, cream, lentils, or other beans, e.g. kidney or white beans or chickpeas)
- A slice of pizza (ask the chef to make it with no cheese)
- Dips, e.g. Baba Ghanouj.
- Some boiled rice with Vegetable Salona (fill three quarters of the plate with vegetables and the remaining quarter with rice)
- Pasta with vegetable and non-creamy sauce (ask the chef to increase vegetable quantity more than the pasta)

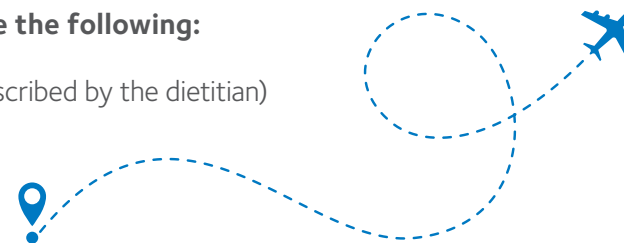
If you like coffee or Karak, keep a bottle of your low protein milk and ask the chef to make your drink using your milk bottle

What should I do if I am travelling?

It is always possible to enjoy travelling while maintaining the low protein diet.

Do not forget to take the following:

- Protein drink (as prescribed by the dietitian)
- Low protein foods
- Medications
- Passport



How to be motivated for the low protein diet?

Try new baking and food recipes using low protein products and ask the Dietitian for guidance.

Try to enjoy meals with the family and friends by sharing new low protein baking and food recipes with loved ones.

Remind yourself and your family that you are doing your best for your health or the health of your child.



What to Do when my Child Gets Admitted to School?

With planning, your child will easily transition to school. Important tips on how to achieve are mentioned below:

1. Who should you inform?

- The principle and admission staff (when enrolling your child)
- The class teachers
- The school nurses
- The canteen staffs

2. What to discuss with those people?

- Provide the staff with a basic understanding of what is HCU and the importance of the medications and the low protein diet
- The condition is not contagious, and it is inherited
- Children with HCU cannot breakdown protein properly so they cannot have high protein food groups
- Eating wrong food will not make them immediately sick but it will have a detrimental long-term effect
- The diet is very specific, and it is calculated by the Dietitian
- The portions and exchanges of certain food that is allowed in the diet
- You must be informed in case your child has eaten food that is not allowed

3. Food and protein substitute at school:

Provide all the food your child will be eating at school

Visit the school canteen and make sure to identify potential food items that are not allowed and highlight that to the staff

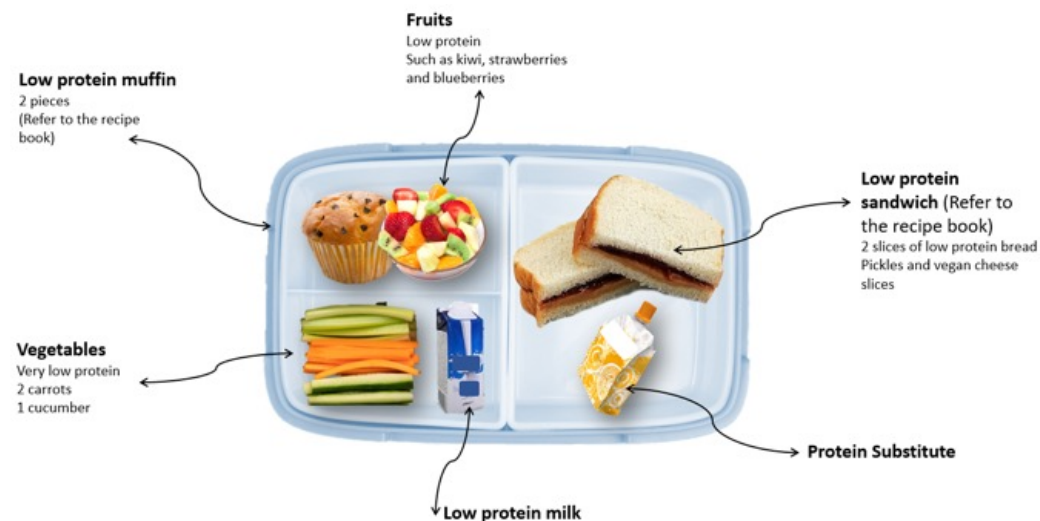
Identify the food that is allowed and highlight it to the staff

In case the school asks for additional information, refer them to your Doctor and your Dietitian for guidance

4. What to discuss with your child?

- Which food are free and which to count
- Bringing home unfinished food to calculate the protein exchanges left
- Not accepting food from anyone at school
- When to drink Low protein milk/formula
- How to explain to the classmates and other friends about the diet

5. Lunch box example



During Illnesses

In periods of short illness such as infections, flu, or cold, it is not a major concern if homocysteine and methionine levels increase if usual treatment (Medication and Low protein diet) is maintained. However, in cases where illness last longer (weeks, or months) there could be a risk of developing health complications, which is why more frequent monitoring is required.

Notice

In case symptoms became worse and persist with vomiting, nausea and oral intake becomes poor, head to the hospital for special management.

For more details refer to the 1g protein exchange, amino acid, and low protein recipe booklets

