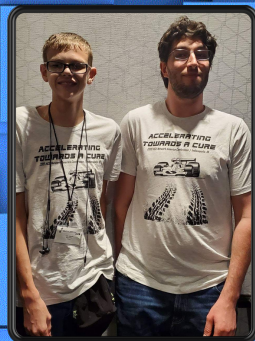


# BACK TO CARE



IT'S NEVER TOO LATE TOO GET BACK ON TRACK

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Thank you to HCU Network America's Patient-Parent Advisory Committee for their assistance in developing the content of this booklet.

### **Disclaimer:**

This booklet does not contain medical advice. Be sure to discuss any medical decisions with your doctor. All information shared by peers is regarding their own personal experiences. Information contained has been scientifically reviewed and approved by the HCU Network America's medical advisory team.

# INTRODUCTION

Welcome back! If you're reading this, you are exploring a return to a plan of care which ultimately will lead to a healthier you. As someone with homocystinuria (HCU), chances are you have felt the huge burden of living with a rare disease. You may also have felt overwhelmed and isolated. Maybe this is why you went off the diet and plan of care in the first place.

You are not alone. We are here with you on your life journey with HCU. We are patients and caregivers who want you to live your best life. This booklet is here to guide you.

Take comfort in knowing that years of medical research have established that there is a management plan for HCU. Most importantly, the plan helps you to achieve and maintain good metabolic control. Before you start to worry about medications, supplements and diet, it is vital to reestablish care with your team of medical experts. Typically, an HCU patient needs to have, at a minimum, a geneticist and a dietitian, both of whom specialize in metabolic disorders. Not sure where to even start? [Click here to see metabolic clinics throughout the U.S.](#) Even if you have to travel a far distance to visit a clinic, it is worth it! Still having trouble? We can help, so email us at [info@hcunetworkamerica.org](mailto:info@hcunetworkamerica.org)

Why is it so important to first reestablish care with an experienced team? Two reasons: (1) the majority of medical professionals are not familiar with HCU, nor do they know how to treat it; (2) all patients with HCU are not the same!

To put it quite simply, you are a rarity within a rarity. In the U.S., approximately 1 in 200,000 people have HCU. And of those with HCU, the treatment plans vary from person to person. What works for one patient might not for another. Some patients have a high protein or methionine tolerance, others low. Some take Betaine, some do not. Some are B6 (pyridoxine) responsive, others are not. Some count grams of protein, and others count milligrams of methionine. So it is vital that you connect with a specialist to establish a plan of care that is as unique as you.

While the plans may vary, the journey is one that you need not travel alone. Together, we are stronger. So start with the very first step: reestablish a plan of care with medical professionals.

# Reestablishing Care



When first returning to care, it is important to visit a metabolic center that specializes in HCU because they will help you set and then reach your HCU goals. They can monitor the amount of homocysteine and methionine in your blood to find your protein tolerance, help you find the best medical food and formula for your needs, prescribe medications and supplements, and provide guidance.

To help you determine if a health care provider is right for you, we suggest asking the provider or their team the following questions and evaluating their answers, during your first appointment:

- How much time do you spend in an appointment?
- Who are the members of the HCU care team? (geneticist, nurse practitioner, dietitian, psychologist, social worker etc.)
- What do you think are the important parts of HCU care?
- How much personal experience do you have with HCU?
- How open are you to new technologies, research, and therapies?
- Who is on call when you are not available?
- Are you available between scheduled appointments, and can I stay in contact with you remotely?

## First Meeting

### Preparing for the Appointment:

- Find out what you'll need before your appointment. Make a call to your new doctor's office and find out exactly what you need to do before getting there.
  - Ask if they want you to prepare a food log. If so, ask how many days do they want you to record your food/beverage intake?
  - Do you need to have lab work done in advance? (They may go over this at the appointment)
  - If able, understand the specifics of your insurance coverage. Does it cover laboratory studies, formula, low protein food and medication? Your clinic should be able to help you with this. If not, email us!  
[info@hcunetworkamerica.org](mailto:info@hcunetworkamerica.org)

# REESTABLISHING CARE

- Be open and honest about what you want and expect from your relationship. That might be a kick-in-the-butt or acknowledgement and support! Either way, be assertive! Your doctor is there to help you, so let them!
- Write down questions for your new doctor before your visit. In addition, try writing down what you are struggling with. Here are some questions to get you thinking.
  - What has been successful for your HCU management and what has not?
  - When is the most challenging time of day?
  - Are there activities during which you struggle to maintain diet
  - What are the social barriers to managing your HCU right now
  - What motivates you?
  - What are your goals?
  - If you are not even sure, determine why you want to improve your care and levels
- Bring everything you need with you. That includes all your insurance cards, and any other paperwork that the doctor's office has requested.
- Ask questions during the appointment to clarify things and to make sure you understand the recommendations or information from your provider.
- Take notes during the appointment so you remember what was discussed.
- Make the adjustments that your doctor has prescribed. It's easy to leave the doctor's office and forget about the changes you said you'd make.

## Schedule Your Next Appointment

Whether with the same doctor, or a new one, schedule your next genetics appointment ASAP so that you don't forget. You can always reschedule it if you need to, but get it on the calendar or odds are you will forget.

### At the Appointment:

- Bring a friend or family member to the appointment. They can be your support system before and after the appointment. They can even take notes!

Initially you will go to clinic more frequently. This process allows your dietitian and physician to help you with an accurate protein tolerance. Once your levels stabilize and your team feels comfortable with your levels they will see you less often.

## Evaluate Your Visit

- Do you like your doctor?
- Do you feel more empowered about your care than when you walked in?
- Were your questions/concerns addressed?

These are really important questions to ask yourself. If the answer is "no", evaluate if there are things that could be done to make the relationship better.

### Patients reestablishing care commonly face a few challenges including the following:

**Challenge:** I know that my homocysteine level is not where I want it to be, and I don't want to feel worse than I already do about it.

#### **Solution:**

- Your doctor's job is to help you better manage your HCU and identify what's not working.
- Ask your geneticist and dietitian for tips/advice that will help you have better success.

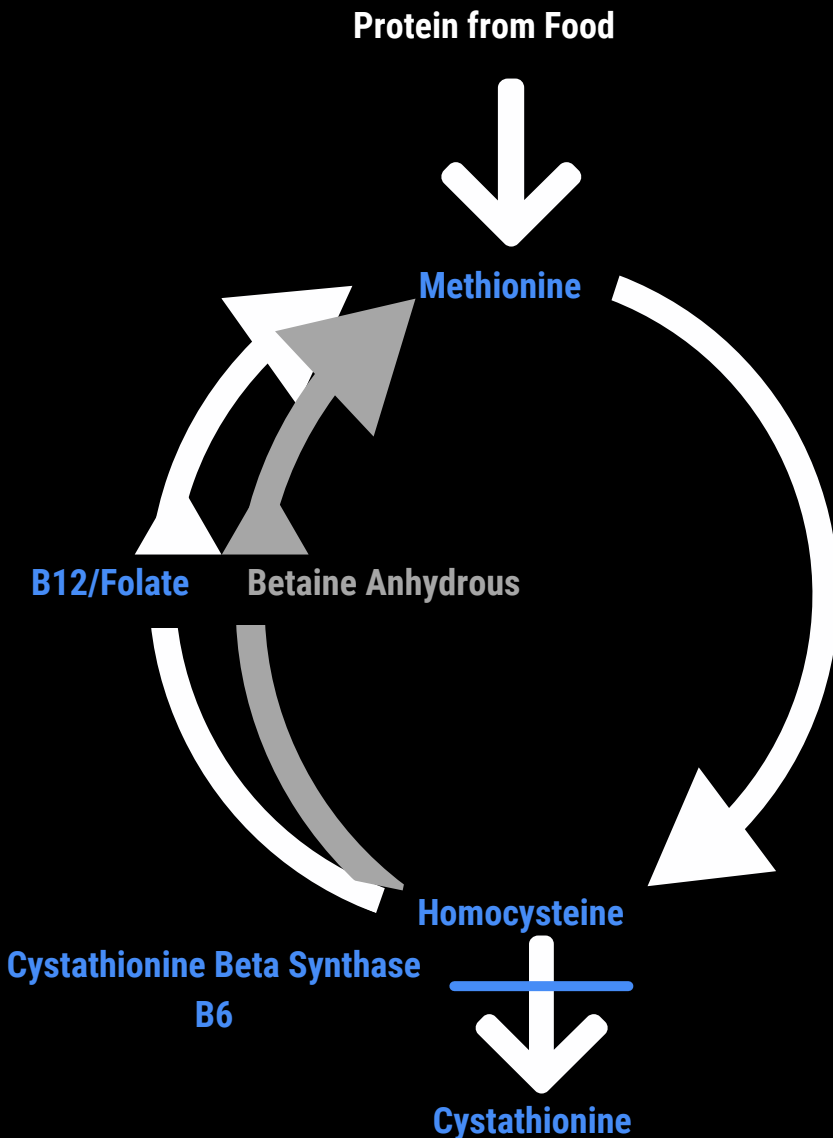
**Challenge:** I don't have enough data for a constructive appointment.

#### **Solution:**

- Understand you have to start somewhere, and it will likely take a few appointments and blood draws to establish a concrete treatment plan.
- Again, it's your doctor's job to help you better manage your HCU and identify what's not working. A good doctor will help you figure out how to get back on track.
- As you know, HCU isn't just about the numbers. You should be able to have a productive appointment with your doctor, even if you're lacking a large amount of data. Talk about what is and isn't working – **be honest!**
- Usually the first meeting with a metabolic specialist is a lot of questions about your health and what you do now. If you know something has not worked in the past, clearly tell your providers.

# Reminder:

## What is HCU?

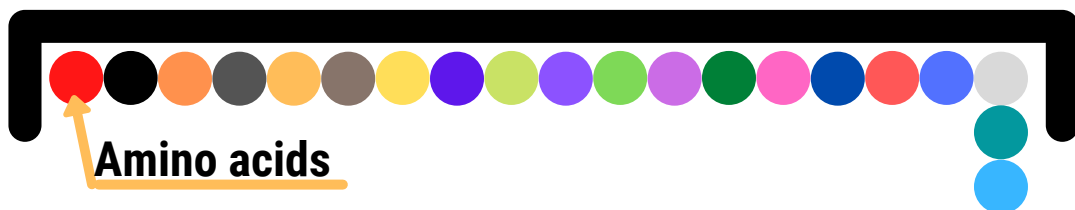




## Words To Know

- **Amino Acids** are the building blocks of protein.
- **Enzymes** help the chemical reactions that break down food and assist with bodily functions at the cellular level.
- **Methionine (MET)** is one of the amino acids/building blocks that makes up protein.
- **Homocysteine (HCY)** is an amino acid that is typically converted to cysteine in the body, but builds up in the bloodstream in enzyme-deficient HCU patients.
- **Protein** is a macronutrient essential for building muscle and tissue in the body, and can be utilized as an energy reserve.

### Protein



Classical Homocystinuria (HCU) is a genetic metabolic disorder where your body cannot metabolize (break down) the amino acid called Homocysteine (HCY) (hō-mō-'si-stə-ēn ) due to the deficiency of an enzyme called Cystathionine Beta Synthase (CBS). Individuals who cannot break down HCY have it accumulate and this also results in elevations in the amino acid Methionine (MET), which is converted to make HCY. MET and HCY are two of the building blocks of protein, which is the long-lasting energy source found in food and used to make more proteins.

MET is contained in proteins that we consume. Normally MET breaks down into the amino acid HCY which is then converted to cysteine. In classical HCU, the conversion to cysteine is defective and the byproduct HCY builds up and has very unhealthy and dangerous clinical effects. High HCY levels are harmful to the eyes, skeletal, vascular and central nervous systems.

# REMINDER: WHAT IS HCU?

## **How does this affect me?**

If your enzymes can't break down HCY completely, it collects in your urine and blood and can cause a variety of symptoms. Most common complications include dislocations of the lens of the eyes; abnormal blood clots; osteoporosis; cognitive deficits; chest deformities, and/ or scoliosis. CBS enzyme activity varies among all patients, so different people experience different effects.

## **Should I feel different?**

Not necessarily, the vast majority of people may feel fine; however, there are very complex processes in the body that may have negative effects in the long term. You are fine until you're not.

## **IMPORTANT Warning for Women!**

Women who wish to start families should be very careful to follow the diet rules before even starting to try to conceive, while trying to conceive, and while pregnant. This is very important for the health and safety of the mother while pregnant.

## **Key Warning Signs and Symptoms Include:**

Patients are born with HCU, however signs and symptoms may develop as early as infancy or as late as adulthood. The symptoms with classical HCU vary greatly, with some affected individuals having only mild signs of the disorder; others may have many different symptoms including some potentially life threatening complications. The most commonly affected areas are: the brain, the eyes, the skeleton, and the vascular system. Due to the symptoms and signs being nonspecific this often leads to misdiagnosis and late diagnosis.

It is important to note that early treatment can prevent the development of symptoms listed on the next page or prevent further complications and worsening of symptoms which are already present.

Speak with your doctor about these potential symptoms and take this HCU Disease Checklist along to your appointment.



## The eyes

- Severe and progressive nearsightedness (myopia) at a young age
- Lens dislocation



## The brain

- Developmental delays
- Cognitive deficit
- Clumsiness
- Psychiatric disorders: anxiety, depression, obsessive-compulsive disorder
- Behavior problems
- Seizures



## The skeleton

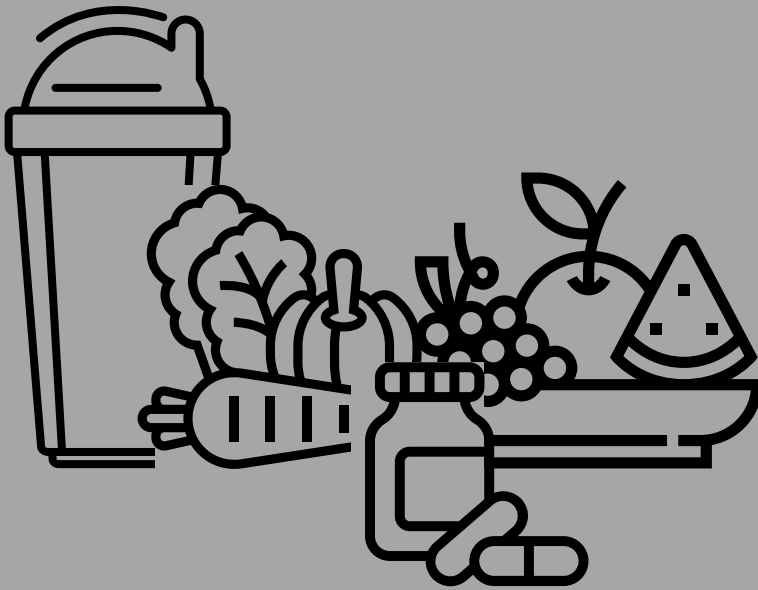
- Marfanoid features (specifically long limbs)
- Excessive growth and thinning or lengthening of the long bones
- Premature osteoporosis
- Scoliosis
- Restricted or excessive joint mobility in the hands
- Abnormally protruding or sunken chest
- Knocked knees
- Highly arched feet



## The vascular system

- Blood clots, especially Deep Venous Thrombosis (DVT)
- Strokes (especially sagittal sinus thrombosis)
- Pulmonary embolism
- Mildly affected individuals may present as adults with blood clots as only problem

# The Diet



### **Challenge:**

I've tried doing the diet and formula cold turkey, but failed.

### **Solution:**

Every clinic has their own method of folding you back into the low protein diet. What has worked for many patients is the following approach:

1. Find a formula you like and get in a routine of taking all of it regularly
2. Eliminate meat – even a bite can mean a lot of protein!
3. Wean yourself off of high protein dairy products: milk, yogurt, cheese – explore low protein alternatives
4. Start supplementing pastas, breads, rice and flours with low protein alternatives. You may find gluten free alternatives at your store that are also lower in protein.

### **Challenge:**

I'm so busy, I don't have the time!

### **Solution:**

Try to plan your meals and meal prep! The biggest issue we have when we are busy is we get pressed for time, don't leave enough time to

prepare something and then end up eating something that we know we really shouldn't be eating. Check out the meal plans and recipes in our [HCU Community Cook Book](#) page of our website for some inspiration.

### **Challenge:**

I'm scared I'm going to gain weight.

### **Solution:**

No one wants to unexpectedly gain 15 pounds. Going back to diet isn't just complicated because of the diet itself, but it can also be frustrating because some people experience unexpected weight gain.

First off, just because something is super low in protein, doesn't mean that it is equally low in calories and that you should binge eat it or even go for seconds. Taking your formula with each meal will help keep you full and from over eating. Try to incorporate high fiber foods with each meal and if you are still hungry, go for more vegetables or fruit.

Don't forget that healthy weekly exercise can keep unwanted weight-gain at bay as well.

## Make Friends With Your Formula and Medication

### You Want Me to Take What?

It doesn't matter if you call it "formula," "milk," or "protein shakes." The goals are the same – protein is a necessary part of anyone's diet, and for people with HCU it just happens to come in a different form: a methionine-free synthetic protein, so you can get the benefit of protein without taking in more MET which would only increase your HCY levels. Your protein supplement is an essential part of your diet, as the amount of protein you are allowed from solid food is likely quite restricted. Without your supplement, your total protein needs are far too low.

Everyone needs to eat every few hours to maintain energy, so it makes sense to have your formula when you eat. Much like food, formula works best when consumed over the course of the day, and not all at once.

**Note:** Following a low protein diet, but not taking formula can leave you with severe nutritional deficiencies and feeling weak. This is NEVER recommended.

### How Do I Get My Formula?

Talk to your HCU dietician about your health and current lifestyle. Your dietitian should be able to send you samples of different types of formula that fit both your nutritional and personal needs. Pick one you like and stick with it.

### I Tried This When I Was a Kid and It Did Not Go Well.

Chances are that you remember the old medical foods and are hesitant to try a new one. Fortunately, medical technology and flavor has improved in the past 50 years. So, pick the formula style that best suits your nutritional needs!

### Types of Medical Foods

Formula has come a long way. For a long time, our only choice was powder in a can but now it not only comes in a can but also in single serving packets, ready-to-drink pouches and even pill form! It comes in different flavors as well. Lots of people also choose unflavored formula and add their own flavoring.



### **Suggested Flavorings**

Kool Aid, Tang, lemonade mix, Mio liquid water enhancer, iced tea, iced coffee, measured amounts of rice, coconut, or almond milk and juice. There are many types of flavorings on the market today. How you make your formula each day may vary, but be consistent in your daily intake.

- Mix exactly how much you need for each meal.
- Mix enough for the whole day and keep it cold in the refrigerator or cooler. Most types can be made ahead of time.
- Keep an extra packet and bottle of water in your backpack, briefcase, or purse.
- Most types of formula cannot be heated up, because they will break down.

### **Tips for Taking Your Medication**

It's very important that you take the medications your medical care team has prescribed for you.

- Use a pill box and fill it once a week to make it more convenient.
- Set an alarm on your phone to remind you to take your medications.
- Mix Betaine Anhydrous with a flavoring that you will like such as Sunny D or different juices.

## What to Eat

People with HCU must avoid foods that are high in protein like meat, fish, poultry, dairy, soy, legumes (dried beans) or nuts. When it comes to fruits and vegetables, there are some that are higher in protein than others. MET is in almost everything except sugar, salt, oil, and water. Thankfully, many fruits and vegetables are naturally low in methionine/protein.

Check out the graphic on the next page to get an idea of some foods that are within your total protein tolerance. Always be mindful of keeping track of your intake so that you stay within your total protein limits per day.

### What If I Miss High Protein Foods?

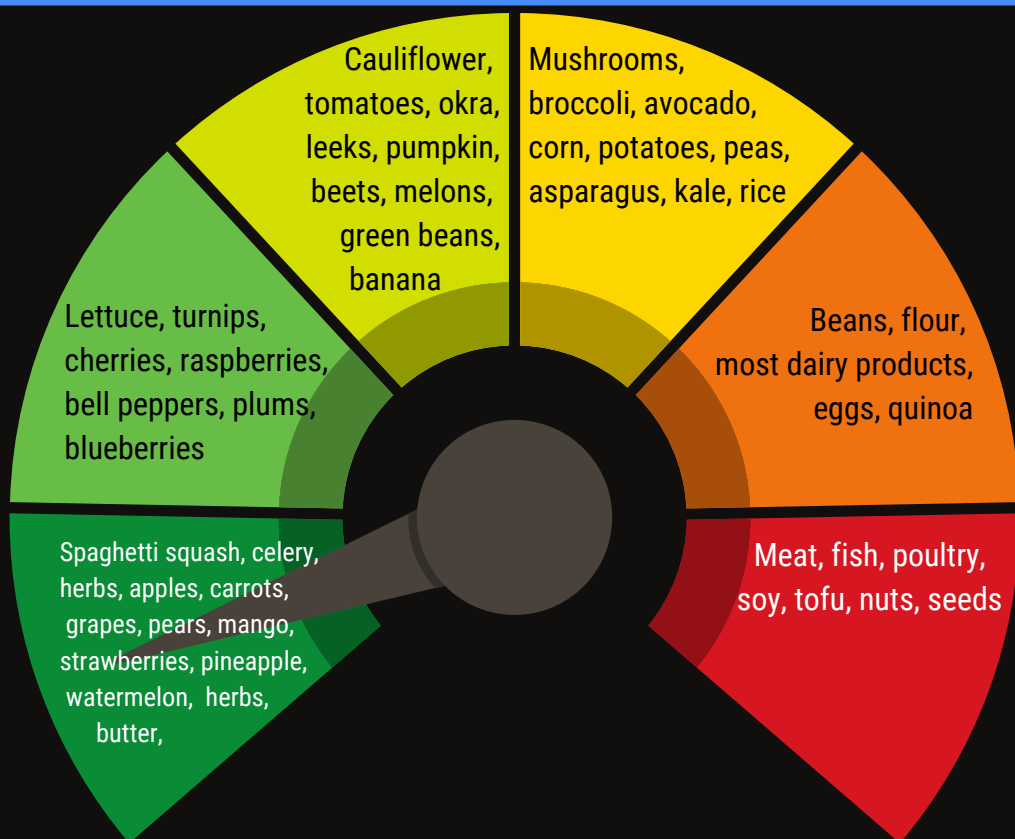
You can also order foods that are modified to be low in protein online, or even in some grocery stores. See page # for a list of suppliers. These suppliers and also most food stores can provide you with a variety of foods lower in protein such as breads, baking mixes, pastas, sauces, desserts, pancakes, waffles, muffins, peanut butter, even cereals and chocolates! You can even buy mixes to make your own burgers, nuggets, and hot dogs. They may be more expensive but provide much more variety. Some states and insurance companies offer help paying for low protein foods.

## Making Good Food Choices

Many things have changed over the past few decades in the world of HCU. We now have low protein cheese, yogurt, meat alternatives, many types of bread, and sweets.

We even have more options in grocery stores now. You can often buy coconut milk yogurt, dairy free cheese, and rice milk in normal grocery stores. Some, but not all, gluten free products are also low in protein and can be found in the grocery store. Be sure to check the nutrition labels before you buy.





Foods shown in the green sections are generally lower in protein per serving. These are great foods to have readily available in case you're hungry and need something low to fill up on.

Foods in yellow tend to have a higher amount of protein in them and should be carefully measured and eaten in moderation. Foods in this section can vary greatly in grams of protein per serving so always be sure to weigh or measure your servings.

Foods in red should basically be avoided if possible. They are very high in protein and could cause your homocysteine level to rise if added to your daily food intake.

**Note:** The protein count will differ for foods when they're cooked, from when they are raw. Think about a baked potato vs potato chips. Brands can also vary.

Small Changes

It will probably be difficult to make huge changes in your lifestyle, but you can start with these lower protein options:

Higher Protein	Lower in Protein
<ul style="list-style-type: none"><li>Bananas</li><li>Creamy soups</li><li>Ground beef</li><li>Mashed potatoes</li><li>Yogurt</li><li>Milk, half and half</li><li>Spaghetti</li><li>Desserts</li></ul>	<ul style="list-style-type: none"><li>Apples, grapes, berries</li><li>Vegetable Soup</li><li>Chopped and seasoned mushrooms or jackfruit</li><li>Mashed cauliflower</li><li>Coconut milk yogurt</li><li>Rice milk, non-dairy creamer</li><li>Bean Thread Noodles, Rice Noodles, Zoodles</li><li>Coconut milk ice cream, sorbet, oreos,</li></ul>

How to Count Protein in Foods

Keep a diet record of EVERYTHING you eat. Look at the example of a breakfast below. Don't be afraid to tell your clinic what you are really eating. They are not there to judge you and will be able to help you better if they know where you are really starting from.

Monday - Breakfast

Quantity	Protein	Food Description
1 Sachet/pouch	0	HCU Express 15 formula powder mixed with lemonade
1 cup coffee	0.2 g	Coffee
1 tbs	0.2 g	Liquid Coffee Mate, original
1	0	Apple
2	2 g	Vans Gluten Free Waffles
2 tbs	0	Syrup
Total protein	2.4 g	

If you're eating a packaged food, it will have a nutrition label on the package. You can estimate how much protein is in these foods with a simple calculation.

**It is important to not only look at the amount of protein listed on the nutritional facts, but to look at the serving size that equates to that amount of protein.**

1. Check the serving size. This food is intended to be eaten in 1/2 cup servings.
2. Now look down to Protein. This says 1 gram of protein for each ½ cup serving.
3. Multiply the number of servings you eat by the protein per serving to get grams of protein per serving. (number servings you eat) X (protein) = (total protein)

# Nutrition Facts

1

Serving Size 1/2 CUP (85g)

Servings Per Container 16

Amount Per Serving

**Calories** 140

Calories from Fat 25

% Daily Value\*

**Total Fat** 3g

**4%**

Saturated Fat 2g

**9%**

Trans Fat 0g

**Cholesterol** 10mg

**4%**

**Sodium** 30mg

**1%**

**Total Carbohydrate** 27g

**9%**

Dietary Fiber 0g

**1%**

Sugars 24g

2

**Protein** 1g

Vitamin A 2%

• Vitamin C 0%

Calcium 6%

• Iron 0%

\*Percent Daily Values are based on a 2,000 calorie diet. Your daily values may be higher or lower depending on your calorie needs:

		Calories	2,000	2,500
Total Fat	Less Than	65g	80g	
Saturated Fat	Less Than	20g	25g	
Cholesterol	Less Than	300mg	300 mg	
Sodium	Less Than	2,400mg	2,400mg	
Total Carbohydrate		300g	375g	
Dietary Fiber		25g	30g	

Calories per gram:

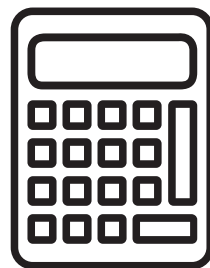
Fat 9 • Carbohydrate 4 • Protein 4

## Other Options for Tracking Protein Intake and Helping Make Healthy Food Choices

Growing up you may have had a cute little chart to track protein intake, which helped foster your independence. Maybe your parents didn't feel so good with that, and they kept a paper diet log. Long gone are those days of losing track of the notebook or paper where your diet log was kept or having to go hunt down the nutrition information to add it into your paper diet log. Now, there are numerous apps to help not only find the protein content of foods, but also track it!

### Things to Consider:

There are many options out there if you have decided to use an app to help you figure out the protein content and log what you are eating. The most important thing to consider is what is best suited for your lifestyle.



**Reliable nutrition information:** Is the nutrition information reliable? Can anyone upload and share nutrition information, or is it coming from a reliable source?

**Rounded vs. Unrounded:** Depending on your protein tolerance, you may be fine with accepting rounded protein – but if you have a lower tolerance, those 2/10th of a point start to add up!

**Ability to export your diet log electronically:** Part of the great wonders of tracking apps is being able to export your diet log to your dietitian.

**Barcode, no barcode?** Some apps now let you scan a food item – this allows you to scan an item and be on your way – one known risk is sometimes the information is inaccurate and regularly rounded.

**Build/modify recipes:** Many apps let you build recipes and modify them as you see fit – this lets you experiment or tweak things to fit your daily allowance.

**Create new entries:** Can't find a food item that you regularly eat? Many apps let you add new food items to your entries.

## Low Protein Tracking Apps

Many low protein food and formula companies have developed apps to help track protein. Here are a few of our top rated items.

### How Much Phe:

Don't let the name fool you, How Much Phe is for any metabolic disorder that requires a low protein diet. How Much Phe allows you to:

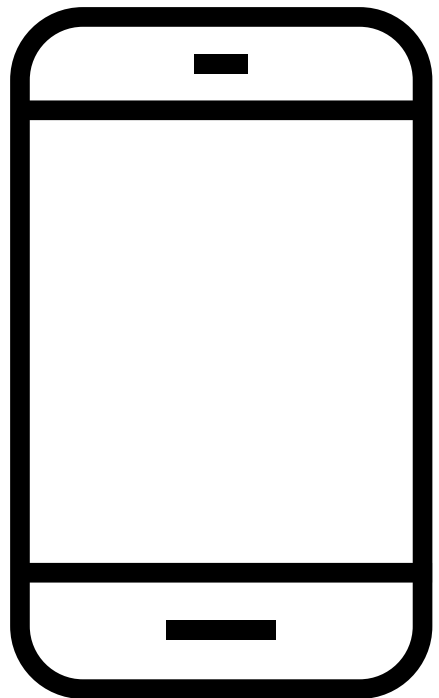
- Web-based, but can be saved as an icon to your phone.
- Find the unrounded protein in over 7,000 items and growing
- Track unrounded protein, or exchanges.
- Add custom foods.
- Build/Modify recipes.
- Meal plan ahead for a day, week or longer.
- Export your diet log.
- <https://howmuchphe.org/>

### HCY Metabolic Diet:

- Web based.
- Syncs with data given to GMDI Metabolic Pro Database.
- Daily nutrient tracker.
- Add your own food or recipe.
- Exports your diet records.
- Allows you to track methionine and protein.
- <http://www.mdapp.org/>

### AccuGo for HCU:

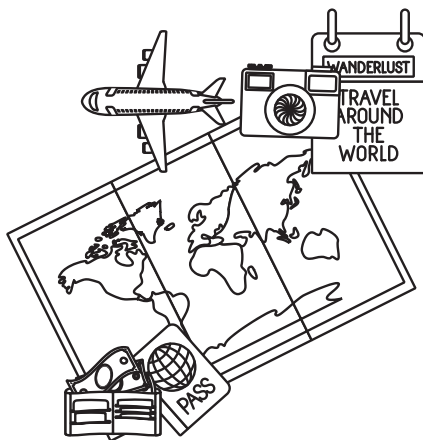
- Only available for iPhone.
- Can work offline.
- Can track protein or methionine.
- Copy/move and delete entries with ease.
- Allows you to menu plan.
- Information based upon USDA based food items.
- Ability to export daily food records.
- <https://apps.apple.com/us/app/accugo-for-hcu/id955040363>



## On the Go with HCU

Having to go out to eat can present a challenge to patients with HCU. It can be tough to access nutritional information and serving sizes, making counting grams of protein/milligrams of methionine feel more like picking lottery numbers than an attempt at science.

When possible, try to limit eating foods with high levels of protein. Instead, opt for vegetables, fruits, salads and other foods that you know are naturally low in protein. Lastly, know your staples. Choose a few foods that are usually available, and try to find out the exact protein counts.



## Low Protein Restaurants: There is an App for That!

It's inevitable that at some point your friends will ask you to dinner, or you will be on vacation and need to grab some items to get you by. For this reason, we recommend a one of a kind app called Go Low Pro.

- Focused on you: Uses location to find locations near you. Filter by restaurant/store/brand to easily find what you need.
- Designed for low-pro: Gluten free is great, vegetarian is fine, but this app is designed JUST for low-protein diets to manage metabolic disorders.
- Brands you know: seeded with products by the members of the PartnershipforPKU.org the app will help you find products your low protein diet requires.
- Powered by you: Share new restaurants and stores complete with product details and pictures. The more you use the app, the better it gets for everyone!
- Traveling? Save room in your luggage: search by postal code and find locations near your destination to pick up low-pro supplies.
- <https://golowpro.org/>

# Use Your Support System



# USE YOUR SUPPORT SYSTEM

## Reach Out

Reach out to others who were diagnosed as adults, or who have navigated coming back to diet. A little support goes a long way. With social networking websites and mobile phone apps it is getting easier to meet other people like yourself. On Facebook it is as easy as searching for the word "HCU", or "Homocystinuria" in the search bar at the top of the page and saying hello to people who show up or joining some of the many HCU groups listed. Many states and regions also have official PKU and Allied Disorder support groups. Your clinic may be able to help you find them. Ask your clinic to put you in touch with other people with HCU and keep you informed about upcoming events. HCU Network America has a list of support groups on their website. Be sure to subscribe to HCU Network America's newsletter, the HCU Herald, to stay up to date on events and new resources.

Reach out to HCU Network America to join one of their Virtual Meetups or be connected with a Mentor!

## Create Support

Support is so important. Your family and close friends, peers or co-workers can provide a source of support. You don't need to tell everyone, but try to find a few people you can turn to when you need a friendly ear or who can learn some basics and help you. Your parents may remember recipes from when you were a child or tricks they used to help you remember to drink your formula.

For those who you invite to this journey, set clear boundaries on what they can help.

### DO

- DO ask me questions about HCU when you are curious.
- DO treat me the same way you treat your other friends.
- DO respect my feelings about HCU.

### DON'T

- DON'T ask me "should you be eating that?"
- DON'T ask me if I'd like "just a bite" of something you know I can't eat
- DON'T look at/comment negatively on my diet.
- DON'T comment on how bad my food or formula may smell.



## Stay Motivated, Not "Perfect"

Many people with HCU feel like they should always have blood levels of homocysteine readings in their target range, and become frustrated when, despite their best efforts, they don't reach their goals. This doesn't have to be a source of stress if you accept the fact that good metabolic care doesn't mean being perfect. Sometimes the elevated level is out of our control, like if you are coming down with a cold. If you forgive yourself for the occasional higher level, you'll be relieved of the stress associated with trying to achieve perfection. You'll likely reap more rewards from this approach in the long term.



## Back to Care Impacts More Than You!

If you have been off diet for a long time, getting back on track may come as a surprise and may be difficult for your family and friends to process. It's not that they don't want you to be healthy, but they likely have become accustomed to you eating similarly to them. Sudden changes in what you were able to eat vs what you are able to eat can be just as difficult for them to process.

### Example:

Your family is celebrating by going out to eat; they pick an upscale steakhouse and ask if that is okay with you. Before you wouldn't blink twice about ordering a steak with its accompanying sides, now your only choice is to get a salad and baked potato. You convey to them that your options are limited and you'd prefer to go somewhere else. They get upset, leaving you left out and make you feel like they don't understand and aren't being supportive. Try to remember, this is an adjustment for both sides and it will take some time to find compromise and routine in what is otherwise a standard scenario.

These situations will make developing a support network that much more important to your success!

## LETTER FROM A PATIENT

It's not that I ever made the intentional decision to go "off diet", it's just that I never really was on diet to begin with. There were times where I was closer to being on diet though, than others, but it still was a far cry from how the diet really should be done. Why you may ask was this the case, well let's start from the beginning.

### **Diagnosis/ Middle School**

In 1995, at the age of 10, I was diagnosed with homocystinuria, shortly after my younger brother was diagnosed. We were trialed on B6, then the addition of betaine, but it was finally decided we needed to follow the low protein diet and formula. We were told to aim for 15 grams of protein from food and to take our formula, which at the time was Hominex-2. We weren't really given much advice or education beyond that. We struggled for a very long time with the diet – in middle school my diet was pretty much sugary cereal, Cheeto's, Budding products (a company that sells very thinly sliced meats so they are 1 g per slice) and pop (Midwest term for soda) – I could fill up and not go over my protein. It was easy to turn to prepackaged foods because the protein content was listed – vegetables and fruit didn't come with labels, so they were this mystery of unknown protein content. Formula on the other hand was a much larger battle – it tasted awful no matter what we did to it. We mixed it with pudding, apple sauce, Jell-O, smoothies – you name it, we tried it, and I was not having it. In middle school though my aunts cornered me and told me that if I didn't take my formula they were going to either force my mom to get me a feeding tube or call Child Protective Services. The thought of a feeding tube or being removed from my family scared me into compliance with my formula.

### **College/Working**

The years went by and having finished my associate degree, it was time to transfer to a 4-year university. As college progressed, my class load became heavier, and I worked more to offset the cost of the classes. I would work 3-12-hour days during the week, cram my classes and assignments into the other 2 days, then work another 16 hours on the weekends. With little spare time, I found myself eating out quite often. When I was with friends between classes, if they offered me food, I would take a bite of a sandwich (with meat) or eat a slice of pizza with cheese. I was so busy that I wasn't thinking of the consequences. By the time I graduated college I was eating regular cheese, bread, pasta and because Hominex doesn't travel well, I was taking it less regularly. Most days I was lucky to finish half. My last semester of college I

ended up having emergency gallbladder surgery. I no longer could take my Hominex because of the large caloric load. Our clinic was able to get HCU Express 15 covered – it really turned out to be a blessing because it was a lot more convenient; it was a much smaller volume and came pre-mixed in individual servings. With the switch, I was able to get in two of the three required servings per day.

In December 2007 I graduated college, but my crazy schedule did not stop. I was motivated to find a teaching job, so I applied in multiple districts as a substitute teacher. Subbing didn't pay very well, so I also had a job in the evenings and on weekends. Similar to my time in college, I was working so much it left little free time. When the new school year started up and I hadn't found a teaching job, I decided to find something that was more consistent and accepted a job teaching preschool in a daycare facility. The hours were consistent, the pay was better, but it would still require me to drive a long way and I'd still need to work on the weekend. Not too long after starting the hours changed. I found myself having to leave home at 5:30 a.m. to get there by 7 a.m. My workday would not end until 6:15 p.m. It made for very long days and I knew something had to change. After a year of working at that location, I accepted a job transfer to a center closer to my home. It was a small pay cut, but I also got back 2.5 hours a day of free time that I had previously spent driving. At this point, I had fallen so deep into my bad food habits, it never even crossed my mind to use that time to reevaluate what I was eating and cook healthier meals.

## **The Blood Clot**

In September 2009, I started having pain in my hand and arm. I went to a clinic and the first doctor told me it was a hang nail. I knew it wasn't a hang nail, but wasn't sure what it was, so I just dismissed it and went on with life. A week later my hand turned blue, then white and very cold – I knew that it wasn't normal so back to the doctor I went. This doctor agreed it wasn't a hang nail, but wasn't sure what was wrong, so he referred me to a hand specialist. The hand specialist immediately recognized it was a blood clot and referred me to pulmonology. I called to make the appointment with pulmonology and they said they couldn't see me for two weeks. It was at that point I finally called my genetic counselor. I told her what was going on and she was flabbergasted and told me to find a ride and come to the hospital that evening. She told me they would have a specialist there to meet me that had expertise in homocystinuria and could address this.

## LETTER FROM A PATIENT

I went to the hospital and met the specialist in the ER. I spent a week in the hospital. I wasn't allowed to do much of anything. If I wanted to eat, I had to have someone feed me. If I had an itch, someone had to scratch my itch for me – they were so afraid any movement would dislodge the clot and cause it to be fatal. After surgery to remove and break up the clot, I had to spend a couple more days there to ensure the left-over pieces didn't cause issues. While in the hospital bed recovering, I realized how lucky I was and that I needed to change my lifestyle.

### After

After the blood clot, I had to be monitored closely by Hematology. I felt like I was always at the lab. They did little though, other than prescribe blood thinners, to address the real heart of the issue – my diet.

Two weeks after being released, I received an invitation to a cooking class hosted by the PKU Organization of Illinois. In one cooking class I learned more about the low protein diet, than I had acquired in the 14 years I had been diagnosed! It felt like there finally was some real education and advice that I had lacked for so long. I quickly started to change my diet. Between the hospital and cooking class, I had already stopped taking bites of things like a burger or sandwich and stopped eating regular cheese. After the cooking class, I quickly made the switch from regular breads and pasta to the low protein versions.

I continued going to events put on by the PKU Organization of Illinois, and at one of their meetings I met Malathy, owner of Taste Connections. I even traveled to Michigan and Iowa to attend her cooking classes. I finally felt like I had options that were not just low protein, but also tasty! I later found Cook for Love and that opened my options even further! I don't think I've ever made a recipe from Brenda Winarski, founder of Cook for Love, that wasn't anything but delicious! I loved cooking before, but now I could cook for myself knowing these recipes were not only delicious, but low in protein.

Throughout the diet transformation, I still struggled with my formula consumption. It just never tasted good and was always clumpy. Then in 2011, I met my now husband. His passion for fitness rubbed off on me, and I started working out, but struggled with weightlifting and extreme muscle fatigue. It would take over a week to recover

## LETTER FROM A PATIENT

from a work out - even after working out regularly for a few months. He had always been great about helping me with the food part of HCU but hadn't really taken much of an interest in the formula portion. It wasn't until he put two and two together that he realized my body wasn't getting enough energy to recover because I wasn't drinking enough formula. He's the one who actually got me to start taking my full dose of formula. Once I started taking the full dose, muscle recovery wasn't really an issue anymore. I was able to quickly lose weight and build muscle.

It's been almost 12 years since my blood clot, and I haven't had any events since. I still struggle with motivation to take my formula but have learned subtle cues that my body needs it – such as brain fog, headaches, fatigue – something that was just a constant part of my life before. I get in food ruts, but then I realize it's time to look at vegetarian/vegan recipes and start trying to adapt them. When I first started my journey back to diet my levels were over 150. The more I cleaned up my diet and the more adherent I was with my formula and betaine the better my levels became. My total homocysteine now is typically 17-20. Being consistent with the diet provides me the best health outcomes and allows me the opportunity to be present for my husband and 2 year old daughter. I'm thankful that I had a second chance and was provided the guidance and education to make my way back to diet.

- Danae' Bartke



Pictured Left to Right: Danae's husband Matt, their daughter Dana, and Danae'

## Low Protein Food Companies

- Lil's Dietary
  - 773-239-0355
  - <https://lilsdietary.com/>
- PKU Perspectives
  - 866-758-3663
  - <https://www.pkuperspectives.com/>
- Taste Connections
  - 310-413-6499
  - <https://tasteconnections.com/>

## Food and Formula Companies

- Cambrooke
  - 866-456-9776
  - <https://www.cambrooke.com/>
- Nutricia North America
  - 800-365-7354
  - <https://www.nutriciametabolics.com/>

## Formula Companies

- Abbott Nutrition
  - 800-551-5838
  - <https://abbottnutrition.com/infant-and-new-mother#metaboli>
- Mead Johnson Nutrition
  - 800-457-3550
  - <https://www.hcp.meadjohnson.com/products/metabolic-products/>
- Nexus Patient Services
  - <https://nexusmedicalnutrition.com/home>
  - 833-875-0200
- Vitaflo
  - 888-848-2356
  - <https://www.vitaflousa.com/>





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