



HCU Network America



Classical Homocystinuria: A toolkit for managing cystathionine beta synthase



HCU Network America

Provided by HCU Network America
www.hcunetworkamerica.org

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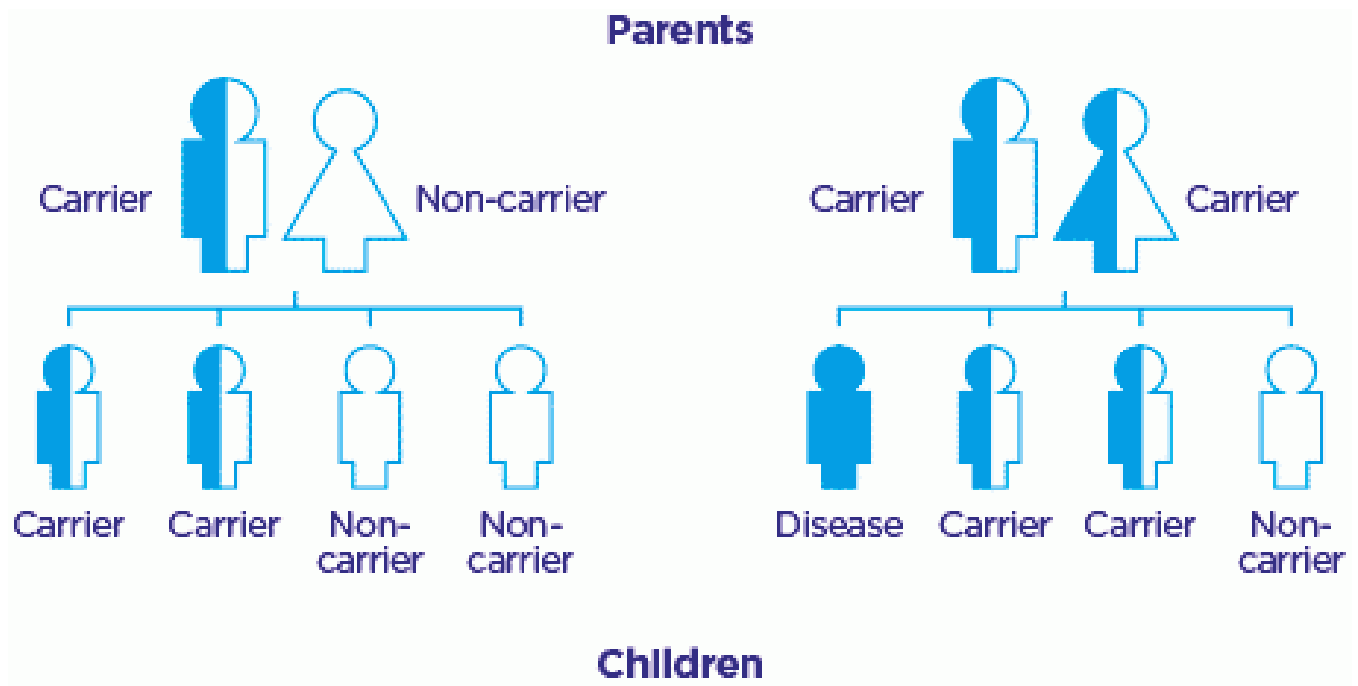
Part 1: What is Homocystinuria?

What is HCU?

Homocystinuria (HCU) is a congenital disorder affecting the metabolism of amino acids.

Classical HCU is caused by the mutation of a gene that codes for Cystathionine Beta Synthase (CBS), the enzyme that metabolizes homocysteine into cystathionine, another amino acid. The absence of CBS prevents this reaction and causes elevated homocysteine, which can cause ophthalmic, circulatory, CNS and skeletal issues. If identified at birth, early treatment can enable normal development and helps decrease the chances of damage.

Classical HCU is an autosomal recessive disease, meaning each parent needs to be a carrier and contribute to the defective gene in the child. Statistically, 1 out of 4 children of carrier parents would be expected to have the disease, with 2 out of 4 becoming carriers.

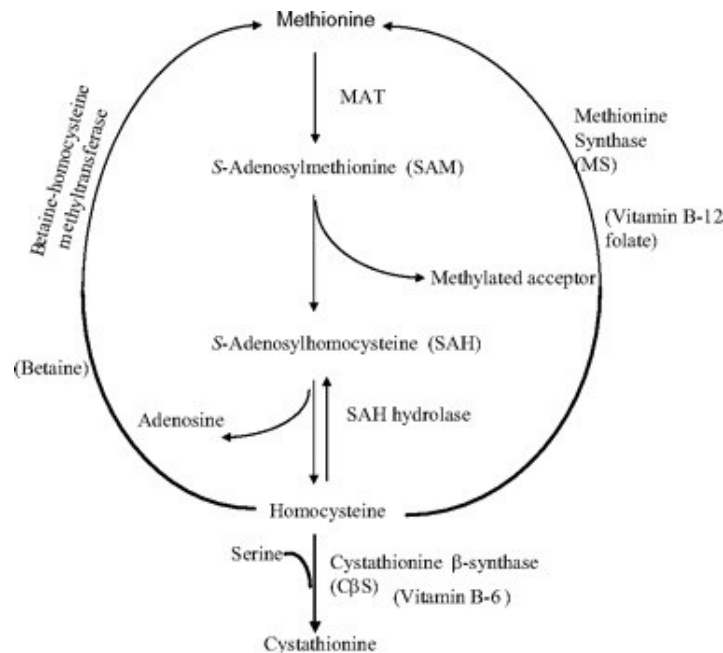


What is Methionine?

Amino Acids like methionine are the building blocks of proteins; methionine is an essential amino acid.

This means that the body cannot make it on its own methionine and therefore it must be provided through food. Methionine is critical in making common proteins.

Homocysteine is also converted back into methionine. Consuming methionine can be problematic for those with Homocystinuria.



How Common is HCU?

HCU is estimated to affect 1 in 200,000-355,000 people in the US, but higher in some countries.

Ireland, Germany, Norway and Qatar have some of the highest incidences of Homocystinuria.

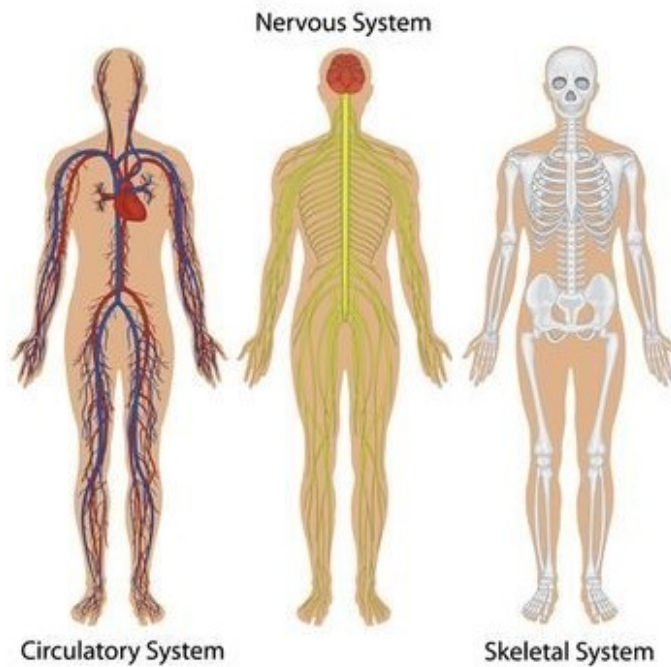
Ireland 1:65,000

Germany 1:17,800

Norway 1:6,400

Qatar 1:1,800

What Areas of the Body are Affected?



Circulatory System

Strokes
Blood Clots
(B6 responsive patients may be asymptomatic or only suffer adult onset thromboembolism)

Central Nervous System

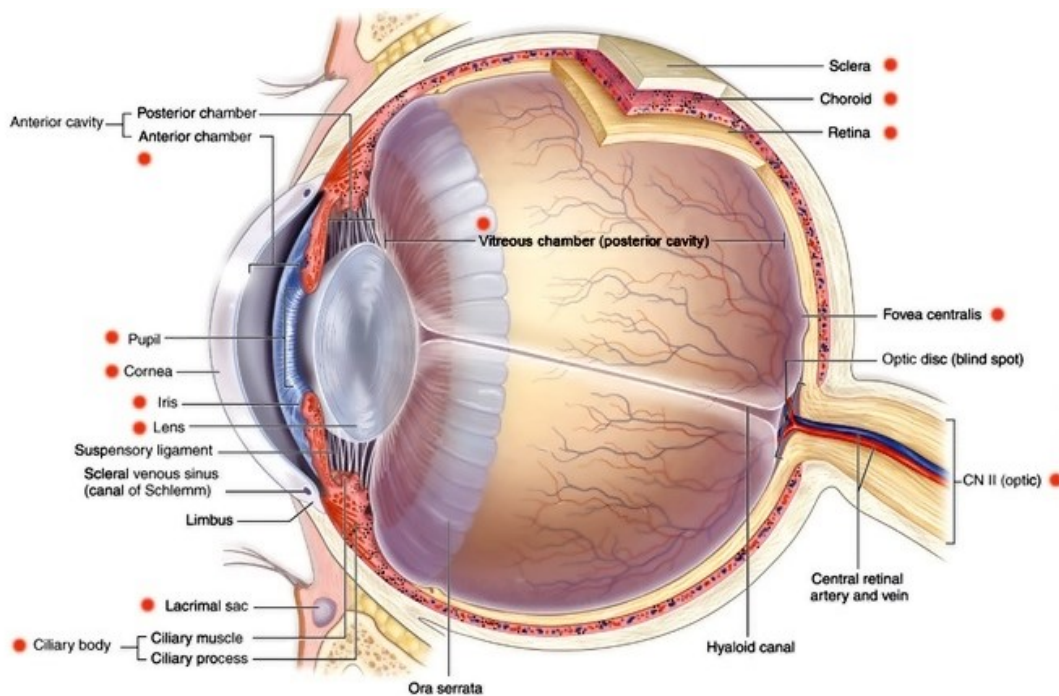
Psychiatric & Behavior Disorders
Learning Difficulties
Intellectual Disability
Movement Disorders
Seizures

Skeletal System

Excessive Height
Long Limbs
Osteoporosis
Bone Deformities
Scoliosis
Knock knees
Pigeon Chest

Eyes

Lens Dislocation
Nearsightedness (Myopia)



How is HCU Diagnosed?

All 50 states screen for HCU at birth.

Newborn Screening

Since 2007 all 50 states have screened for HCU at birth via newborn screening. Unfortunately though, at least 50% of cases are not picked up by newborn screening. These 50% of patients are often but not always those who have the milder, vitamin B6 (pyridoxine) responsive or partially vitamin B6 responsive form.

- The newborn screening test uses a heel prick to measure methionine levels – if high, homocysteine levels are then measured

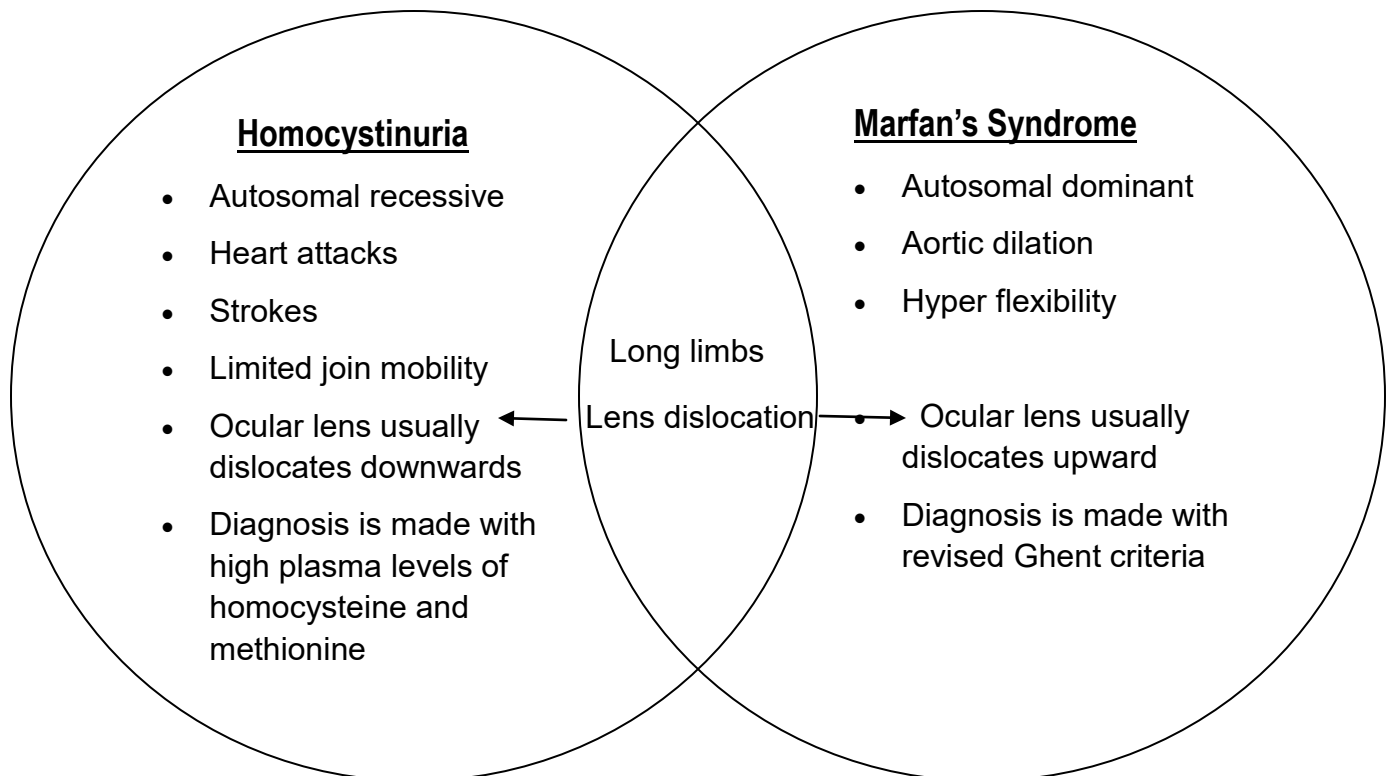
Late Diagnoses

- If missed by newborn screening, HCU can be diagnosed later due to clinical symptoms. It takes an average of 4.5 years from first symptoms to receive an accurate diagnosis.
- By 10 years of age, without treatment 55% of B6-responsive and 82% of non-responsive will have lens dislocation
- By 15 years without treatment 12% of B6-responsive and 27% of non-responsive will have a thromboembolic event.

When testing for HCU doctors should use the plasma total homocysteine test since testing free homocysteine is unreliable

- Patients should not be taking a multivitamin or vitamin B6 prior to the blood test.
- Plasma should be separated promptly

Homocystinuria (CBS Deficiency) can be confirmed by mutation analysis or CBS assay. Because HCU shares many similar skeletal and eye features with Marfan's Syndrome, people with HCU are mistakenly diagnosed as having Marfan's Syndrome.



What is the Clinical Presentation?

In a healthy person who is not affected by HCU, plasma homocysteine usually will not exceed 15 (umol/L) of blood. Healthy carriers may have less ability to metabolize homocysteine and have elevated levels that exceed 15umol/L. If you are a carrier with heightened levels, please seek the advice of a metabolic specialist.

Patients on treatment may have levels greater than 15 (umol/L), untreated patients often have levels several fold higher.

B6 (Pyridoxine) Responsive or Not

HCU patients can be broken down into two categories, B6 responsive or non-responsive.

B6 Responsive

About 50% of patients are vitamin B6 (pyridoxine) responsive or partially vitamin B6 responsive. A few patients who are vitamin B6 responsive do not require a low-protein diet, but most still require some restriction of protein and/or the HCU formula. If patients cannot reduce their homocysteine levels below 50 umol/L with B6 alone, they may require the addition of betaine to their daily regimen.

B6 Non-Responsive

The other half of patients are considered B6 non responsive. B6 non-responsive patients are usually on a low-protein diet, often with betaine added, and require specialty low-protein medical foods and supplements of other amino acids. The goal for these patients is to reach levels below 100 umol/L, though some physicians will recommend lower levels if they can be reached safely.

Prognosis

Early diagnosis and adherence can prevent the onset of the symptoms.

Adherence to the medication and diet must be constantly monitored. If the homocysteine level is kept below 70-100umol/L, cognitive and developmental delays, as well as the chance for blood clots and strokes can be greatly minimized. Some clinics may aim for lower levels.

Monitoring

Frequency of clinic visits and lab tests are dependent upon severity, treatment and age.

Total homocysteine and methionine values should be monitored and discussed at each visit.

In addition, patients should periodically have a folate, vitamin B12, full amino acid panel and bone density scan.

What Causes High Levels

Multiple factors can play a role in patients' homocysteine and methionine concentrations.

- Endogenous Inborn Errors of the Metabolism
- Excessive protein intake, resulting in increased methionine
- Lack of CBS enzyme activity
- Vitamin B12 or folate deficiency
- Methylation disorder (e.g MTHFR)
- Sickness or stress (any kind), resulting in catabolism – the breakdown of muscle that releases amino acids into the blood for energy

Part 2: HCU Nutritional Guide

Diet and Medication

Medication

Vitamin B6 is given to B6 responsive patients to stabilize any remaining CBS enzyme. If folate (B9) or vitamin B12 (Cobalamin) levels are low, elevated homocysteine may result, so many patients take low doses of B12 and folate.

For patients who cannot reduce homocysteine to the targeted range, they may require the addition of Betaine, a supplement that helps convert homocysteine back to methionine. Betaine dosage depends on the level of (if any) active enzyme, height, weight and age.

Your healthcare provider may have you add other vitamins or medication to ensure your health is optimized.

Low Methionine/ Protein Diet

Methods for calculating intake vary among metabolic clinics. Some recommend that patients who require a low-protein diet count methionine intake from food (in milligrams), while other clinics recommend that patients count overall protein intake (in grams). The intake goal will vary depending on the patient's homocysteine and methionine levels. Both methods require careful measurement of food. Patients will get most of their protein from fruits, vegetables and synthetic low-protein foods, which include low-protein pasta, bread, rice, dairy, egg, dessert and meat substitutes.

HCU Medical Formula

An important component for success to those on a low-protein diet is HCU medical formula. Without HCU formula, it can nearly be impossible to meet total protein requirements for normal growth and development. Formula provides all amino acids (except methionine) and the majority of vitamins and minerals a person needs while on a low-protein diet.

*Please see our Medical Formula and Low-protein vendor list for additional resources

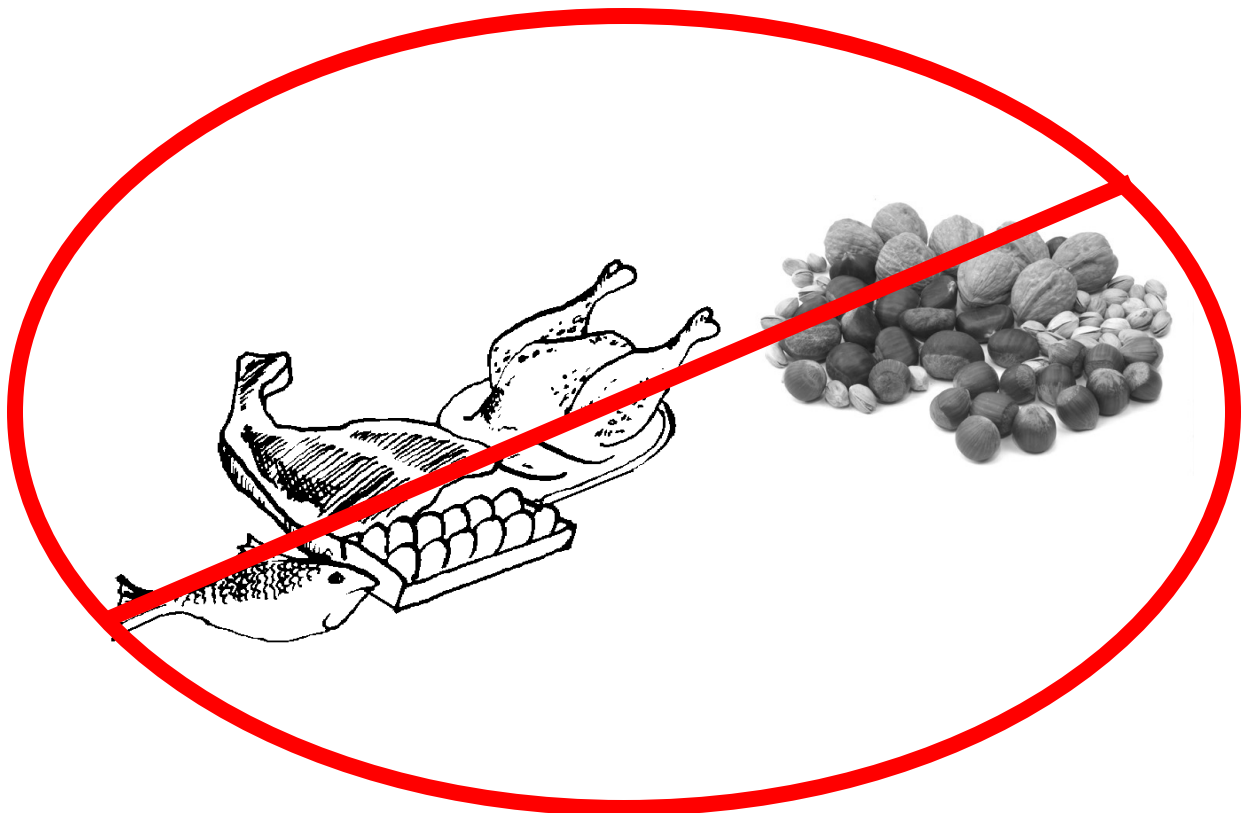
Basic Rules of the Diet

The dietary regimen has some ground rules, the most important being to keep plasma Homocysteine levels within safe limits.

Methionine is found in all natural proteins. Because methionine is an essential amino acid, everyone needs some methionine in their diet. For patients with HCU, providing just enough methionine to meet needs for growth and/or maintenance is the key. Calories from other foods, including low-protein food/medical formula, are very important and come from the following sources:

- Foods naturally lower in protein, like fruits and vegetables, are recommended as the staple of the diet regime.
- Special low-protein foods such as flour, bread and pasta.
- Measured amounts of regular grains, such as rice and crackers
- Special low-protein food and medical formula
- All patients should be followed by a metabolic dietitian who can help personalize their diet therapy.

Note that the dietary guidelines for HCU do **NOT** include high protein foods such as meat, fish, eggs, soy, nuts and even some high protein grains. Patients must make sure they are consuming adequate amounts of vitamins, minerals, carbohydrates and essential fatty acids for their age, sex and energy needs. Consulting a dietitian will help you find the right medical formula and natural protein combination. With their advice, patients will be able to develop and grow at a normal rate while restricting their methionine intake.



What are the Recommended Protein Levels?

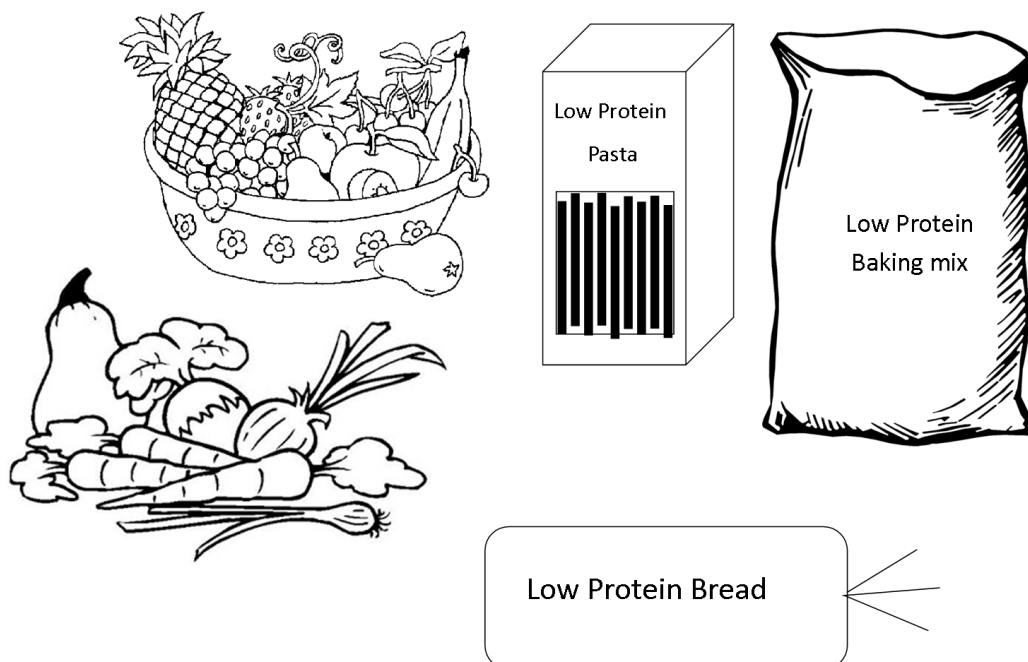
A low-protein diet must always meet individual protein needs. Recommendations for total protein (protein from food, plus medical formula) vary with age and can be between 1 and 3 grams of protein per kilogram of body weight per day.

Work with your dietitian to find the right balance of total protein for you.

HCU Medical Formula



Low-protein Foods



Feeding Babies with HCU

When HCU is diagnosed as a result of Newborn Screening, it is important to begin dietary treatment immediately.

Breastmilk and standard infant formulas contain small quantities of methionine. Breast feeding babies with HCU is encouraged and helps create a strong tie between mother and child. Breastfeeding and infant formula may need to be supplemented with an appropriate HCU medical formula.



Feeding Children with HCU

Children are growing and have high calorie and protein requirements.

A child's methionine/natural protein tolerance will dictate the dietary approach. A diet deficient in total protein would result in poor growth. There must be enough total protein on board to keep the child healthy and ensure growth. This cannot be accomplished without medical food.

Additionally, since methionine is an essential amino acid, it is rate-limiting, which means that too little natural protein from food will also result in poor weight gain and growth. Methionine intake cannot be "too much" or "too little" – it must be "just right."

While watching the protein or methionine content, it's also important to make sure the caloric needs are being met. Insufficient calories could compromise growth and lead to catabolism of protein from muscle, which would release methionine.

The diet should be monitored by a daily diet log and by blood draws. Your dietitian and geneticist will make adjustments from your diet log and your lab results



HCU in Teenagers and Adults

The correct diet allows normal development of adolescents, mentally and physically, helping them lead normal lives.

The teenage and adult medical formula supplements are available in easy packaging to accommodate an active lifestyle.

Amino acid requirements for adults are not greatly different than that of children, but there are other nutrients that are just as important:

- Calcium and Vitamin D: to prevent osteoporosis and promote bone health
- Folic Acid: to prevent arteriosclerosis
- Fiber: for normal digestive health
- Omega 3 essential fatty acids: which strengthen immunity and have neuroprotective effects.

Metabolic clinic visits are less frequent in adulthood than in childhood, but regular monitoring is still important.



Part 3: HCU and Pregnancy

Birth Control and Pregnancy

It's especially important for women with HCU to take the proper precautions before and during pregnancy to ensure they have a safe and successful pregnancy.

As previously stated, HCU comes with an increased risk for blood clots. Estrogen-based birth control, even in the general population, increases the chances for a blood clot. Because of this, it is highly recommended that women with HCU avoid estrogen-based birth control.

If possible, women should consult with their metabolic team before getting pregnant, about their decision to get pregnant. While pregnancy does pose some risk for women with HCU, it can be done in a very safe manner. It is likely you will be advised to consult with a hematologist and maternal fetal medicine specialist.

It is important that you have your metabolic team follow you closely when considering getting pregnant and during the pregnancy itself.

In the case of women who are not B6 responsive, it is also important that you closely follow your prescribed food and formula diet that your metabolic team has prescribed for you. As you progress in your pregnancy, your metabolic team will likely adjust your formula and protein intake to make sure you are meeting you and your growing baby's needs!

Part 4: Tips and Tricks

Don't get hung up on the details - here are a few tips and tricks to help you be more successful.

- Look at the food labels
 - Double check the serving size and protein content for the listed serving.

Nutrition Facts		
Serving Size		
Serving per Container		
Amount Per Serving		
Calories	Calories From Fat	
% Daily Value *		
Total Fat	...g	...%
Saturated Fat	...g	...%
Cholesterol	...g	...%
Sodium	...mg	...%
Total Carbohydrate	...g	...%
Dietary Fiber	...g	...%
Sugar	...g	...%
Protein	...g	...%
<hr/>		
Vitamin A	...%	• Vitamin C ...%
Calcium	...%	• Iron ...%

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

- Generally, foods are weighed after they have been peeled or trimmed and before cooking.

Ingredients in many high protein recipes can be swapped out for low-protein items.

- Eggs –
 - For one egg, use ¼ cup unsweetened applesauce
 - For one egg, use ¼ cup smashed banana
 - For one egg, use ¼ cup low-protein plain yogurt
 - Ener-g® Egg Replacer – follow directions on box
- Milk-
 - Cooking
 - Rice milk – ¾ cup for every 1 cup regular milk.
 - Baking -
 - Almond milk 1 cup for every cup.
- Flour-
 - Low-protein baking mix – follow directions on box
- Cheese
 - High protein cheese can be substituted for low-protein or *some* vegan cheeses.

Eating Out

Having HCU doesn't mean the end of eating out.

People and restaurants are more aware of food allergies and special diets than they have ever been! Keep these ideas in mind when you visit a restaurant or fast food joint.

- Keep a small food scale with you to weigh your items
- Try to view their menu online in advance
- Many restaurants have their nutrition information available online or available if you call
- If you don't see something that suites your diet perfectly, don't be afraid to ask if they can substitute something
 - Example, a Portobello mushroom for a beef hamburger patty
- Some restaurants will let you bring your own low-protein pasta and cook it for you. Call ahead to see if they will cook or heat up your pasta for you



Traveling



Exploration shouldn't be limited by HCU

Prepare yourself and give yourself plenty of time. This will help ensure that your travels will go smoothly and will be enjoyable. Here are some tips to help you on your way.

- Gather a note of medical necessity from your metabolic doctor
 - This note should explain Homocystinuria, what medications, supplements, formula and food items you may be traveling with
- Check out the food scene where you are going; it may be necessary to pack your low-protein foods
- Pack extra food and formula, you never know when your flight might get delayed
- Carry as much of your medication, supplements and food as a carry on. Some airlines will allow an additional carry on if medically necessary. Check with your airline to see if they have such a policy
- Arrive at the airport with plenty of time. Because of the nature of supplements and formula, your luggage will likely get pulled aside and searched. Be patient and show them your letter of medical necessity. They will still check the contents, but it will make things move much smoothly!
- Have a written letter of what should happen in case of a medical emergency. Work with your medical team to complete this.
- Once on the airplane or in the vehicle, make sure you take plenty of stretch breaks to promote circulation.



Part 5: Food and Formula Resources

Medical Formula Vendors

Abbot Nutrition

Abbott Nutrition metabolic medical foods are designed to help meet the specialized nutrient needs of patients who have an inherited metabolic disorder. The formulation and nutrient composition of our metabolic medical foods are based on clinical research findings and recognized nutrient needs for growth and maintenance of nutrition status for infants, toddlers, children, and adults.

Formula currently available for HCU:

Hominex®-1
Hominex®-2

Contact:

<https://abbottnutrition.com/infant-and-new-mother#metabolic>

Mead-Johnson

For more than 50 years, Mead Johnson has been a pioneer in the field of specialized nutrition for inborn errors of metabolism. Today, the company commercializes several metabolic formulas designed for the dietary management of specific conditions, including phenylketonuria, maple syrup urine disease and homocystinuria, among others.

Formula currently available for HCU:

HCY 1
HCY 2

Contact:

Phone: (812) 429-6399

Website: <https://www.meadjohnson.com/pediatrics/us-en/product-information/products#metabolics>

Vitaflo USA

For over 20 years, Vitaflo® has been at the forefront of developing innovative specialized clinical nutrition products for Inborn Errors of Metabolism (IEM), nutrition support and disease specific conditions such as renal disease. Our dedicated experts have over 30 years' experience in producing specialist nutritional products. Working in partnership with world key opinion leaders and patients, Vitaflo's aim is to create products that combine the best of cutting edge research with the lifestyle demands of modern living, ensuring the most acceptable products are available for the patient.

Formula currently available for HCU:

HCU Gel
HCU Express
HCU Cooler

Contact:

Phone: (888) 848-2356

Email: CustomerService@VitafloUSA.com

Website: <https://www.nestlehealthscience.us/vitaflo-usa/inborn-errors-of-metabolism/protein-metabolism/>

Low-protein Foods and Medical Formula Vendors

Cambrooke Therapeutics

Founded in 2000, Cambrooke Therapeutics recognizes the critical nature of managing serious medical disorders through medically-based nutrition therapies. For people with serious unmet medical nutrition needs, it is a daily challenge to find flavorful, easy options that can sustain their diets and maintain their nutritional needs.

Formula currently available for HCU:
Homoactin AA **Plus** Berry

Contact:

Phone: (866) 456-9776

Email: info@cambrooke.com

Website: <http://www.cambrooke.com/>

Nutricia Metabolics

Nutricia is a global health company that leads the development and use of advanced medical nutrition for specialized care. Nutricia products and services transform the daily lives of millions of infants, children, adults and aging seniors. We are dedicated to providing quality formulas and medical foods for the treatment of food protein allergies and rare genetic and metabolic disorders.

Formula currently available for HCU:

HCU Anamix: Early Years

HCU Anamix: Next

HCU Lophlex LQ

XMet Maxamaid

XMET Maxamum

Milupa Hom 2

Contact:

Phone: (800) 605-0410

Email: info@medicalfood.com

Website: <http://www.medicalfood.com/>

Low-protein Food Vendors

Dietary Specialty

Dietary Specialties has earned a reputation for exceptional products for people on low-protein or gluten free diets for more than fifty years. Our unique foods are produced from the finest ingredients under rigid quality control standards to ensure that they are totally appropriate for your special diet. Let us help you discover that being on a restricted diet does not mean sacrificing great taste, variety or convenience. Dietary Specialties' Low-protein foods make managing diets for inborn metabolic conditions like PKU, MSUD and other amino acid disorders easier and much more manageable with the right foods!

Contact:

Phone: (888) 640-2800

Website: <http://dietspec.com>

Flavis

The new Flavis low-protein range contains that all-important ingredient that we believe has been missing from low-protein products available until now... flavor.

Contact:

Email: info.us@flavis.com

Website: <https://shop.flavis.com>

Lil's Dietary Shop

We are the diet food shop. We are a provider of medically necessary food. We offer selections in pastas, frozen foods, cookies, crackers, baking and flour mixes, breads, sauces, cereals, etc. We are continually searching for the special request of our customers. If it is out there, we will find it!

Contact:

Phone: (773) 239-0355

Website: Lilsdietary.com

Promin Metabolics

Promin was set up specifically to focus exclusively on the production of specialized dietary products, placing the needs of people directed to follow a special diet daily at the core of its business model. Currently, over 70 unique products are imported and distributed nationally in the United States and we will soon be announcing alternative delivery options as well.

Contact:

Phone: (240)513-6889

Email: sales@prominmetabolics.com

Website: <http://prominmetabolics.com>

PKU Perspectives

PKU Perspectives was formed to share with all PKU families and those on low-protein diets, the convenience foods we have learned to enjoy. After preparing meals for two PKU children of our own and an adopted handicapped grandchild with PKU for more than thirty years, we have discovered that convenience is most appreciated when you must prepare two meals at a time. PKU Perspectives is a for profit company. We employ many individuals who have a deep interest in providing products for special dietary needs.

Contact:

Phone: (866) 758-3663

Email: Sales@pkuperspectives.com

Website: <http://pkuperspectives.com>

Taste Connections

We specialize in low-protein products such as bread mix, multi-baking mix, etc. breads, cookies, and other baked goods that are suitable for metabolic conditions such as Phenylketonuria, Homocystinuria, etc. We are committed to providing best quality low-protein products to our customers at lowest possible price. In the near future, we will be adding several new low-protein products for your convenience.

Contact:

Phone: (310) 413-6499

Email: tasteconnect@verizon.net

Website: <http://Tasteconnections.com>

Other Notable Food Companies

While these companies do not design things with our community in mind, many of their products will work for those on a low-protein diet. Please check the nutrition labels and consult with your dietician if you are not sure if their products are right for you. Most of these companies can be found in most grocery stores or health food stores.

Better Than Foods

At Better Than Foods, we have a range of healthy and tasty pasta noodles and rice alternatives. Our organic Konjac range is made from organic Konnyaku flour from the Konjac plant. Our healthy ready to eat meals are made from a unique Konjac recipe with no artificial colors, flavors or preservatives. Our latest addition is the Konjac range made from a unique recipe that gives our konjac food a texture far better than regular konjac and without the need of water, so there's no need to drain or rinse.

Contact:

Phone: (855) 691-5900

Email: Jeff@GreenSpotFoods.com

Website: <http://www.betterthanfoodsusa.com/WebV31/>

Daiya Foods

Friends for over 20 years, Andre and Greg share a passion for music, ideation and a plant-based lifestyle. The same passion inspired them to create a delicious dairy-free alternative made entirely from plants.

Their perseverance paid off and Daiya Cheddar Style and Mozzarella Style Shreds debuted in 2009 at the Natural Products Expo West. Consumers raved — and plant-based dairy innovation changed forever.

Contact:

Phone: (877) 324-9211

Email: cr@daiyafoods.com

website: <https://daiyafoods.com>

Ener-g Foods Company

While Ener-g foods does carry some items that are specific to the low-protein community (please see their specialty items for all low-protein items), they have branched out to other dietary restricted communities as well. They carry many baking mixes, breads, and snack items.

Contact:

Phone: (800) 331-5222

Email: customerservice@ener-g.com

Website: <http://www.ener-g.com>

Field Roast: Chao Slices

(We have decided to focus on their Chao slices as their other products do not pertain to a low-protein diet)

Chao Slices! Welcome to Field Roast's newest family member: a vegan coconut cheese alternative seasoned with a traditionally fermented soybean curd called Chao by the Vietnamese. We continue our fascination with blending Asian and European heritages by partnering with a Greek cheese maker and a family in Taiwan who makes the Chao. We bring it all together in Seattle to create Chao Slices.

Contact:

Phone: (800) 311-9497

Website: <http://fieldroast.com/product/chao-slices/>

Follow Your Heart

Follow Your Heart is thriving and still committed to the same ideals and values that we had in 1970. Since our humble beginnings as a small market and café in Canoga Park, we have expanded our passion for great-tasting, healthy foods. We believe that all people deserve access to healthy food, that we should have compassion for all living things, and that we have a responsibility to be good stewards of the environment.

Contact:

Phone: (818) 348-3240

Email: amazon@followyourheart.com

Website: <http://followyourheart.com>

Glutino

Glutino Gluten Free offers an array of baking mixes, snacks, breads, and breakfast pastries.

Contact:

Phone: (800) 845-7286

Website: <http://glutino.com>

The Jackfruit Company

Annie founded Global Village Fruit in 2011 after learning that jackfruit is under-researched and underutilized on a global scale. The largest producer of the world's jackfruit is the country of India, where 75% of the fruit goes to waste largely for lack of processing infrastructure, refrigerated supply chains, and connections between farmers and markets. That's why Global Village Fruit is on the scene, building international supply chains. Knowing this wonderfruit would change the globe, Annie decided to brand The Jackfruit Company as the face of jackfruit foods in the US. We are now the #1 jackfruit brand in retail stores nationwide!

Contact:

Phone: (877) 433-4024

Email: knowjack@thejackfruitcompany.com

Website: <http://thejackfruitcompany.com>

So Delicious

So Delicious Dairy Free has a variety of dairy free products including, Milk alternatives, non-dairy creamer, yogurt and cold desserts.

Contact:

Phone: (866) 388-7853

Website: <http://sodeliciousdairyfree.com/>

Upton's Naturals

Upton's Naturals is an independently-owned natural foods company with a focus on meat alternatives and vegan values since opening in 2006. Dedicated to simplicity and the use of real, recognizable ingredients, the Upton's team takes food that has been around for thousands of years and offers them to consumers worldwide. The company's founder Daniel Staackmann pioneered flavored, wheat-based seitan and has now launched the first-to-market, seasoned and ready-to-eat jackfruit in the United States.

Contact:

Phone: (312) 666-7838

Email: info@uptonsnaturals.com

Website: uptonsnaturals.com

Important Drug Companies

Recordati Rare Rare Diseases (Makers of Cystadane)
Cystadane can be purchased by prescription through AnovoRx

Contact:

Website: <https://www.cystadane.com/>

Phone: (844) 288-5007

Anovo's Website: <http://www.anovorx.com/>

Anovo's Phone: (888) 487-4703

Do you have a suggestion to add to our list?

Email us! Hcunetworkamerica@gmail.com



Low-protein Food and Medical Formula Insurance and Assistance Programs

We realize that food and formula coverage varies greatly by state. For this reason, HCU Network America has contracted Raenette Franco of Compassion Works Medical LLC to help assist you in getting low-protein foods, formula and supplements covered. She will also work with you to find a policy that meets your individual needs if you don't currently have one or are struggling to find one.

Part of our mission as HCU Network America is to increase access for treatments and supplements for Homocystinuria patients. In order to fulfill this part of our mission, we have been in contact with Raenette Franco of Compassion Works Medical, LLC to help us fulfill this part of our goals.



About Raenette

A native New Yorker, Raenette Franco came to New Jersey in 2001 and landed a career as Medical Biller Insurance Specialist/Consultant. Her background consists of hospice and palliative care, bariatric surgery, and other medical specialties geared around insurance coverage. Raenette landed a job within the medical food arena in 2011 as a medical food insurance specialist that captured her heart. Inspired by helping others, Raenette expanded her career as founder of Compassion Works Medical. Raenette received her Certification as a Certified Biller Coder Specialist (CBCS), from the NCCA Accreditation, NJ. Her big heart and deep passion is dedicated to helping patients, dietitians, and physicians avoid the difficult tasks of insurance coverage and reimbursement issues for medical foods/enteral nutrition. Raenette's specialty is working directly with Medical Food coverage for both formula

and low-protein foods.

Mission

Compassion Works Medical has a special mission to change the lives of the rare genetic disease community by supporting a genuine helping hand with insurance coverage challenges. Over the past years at Compassion Works Medical, a blessing in disguise had come across my path – I have expanded my support to all kinds of special diseases that require medical foods and enteral nutrition, including cancer, ALS, TBI, etc. It is truly rewarding and my inspiration grows stronger and stronger to help those people find a way to afford and stay on diet.

“We don't take NO for an answer along with compassion that makes all of us successful”!

How to get in touch with Raenette:

Email: raenettef@compassionworksmrs.com | Phone: (973) 832-4736

Other Assistance Programs

Healthwell Foundation

Each year, more and more Americans are being forced to choose between paying for life-saving treatments and paying for basic necessities like food, housing and utilities – decisions no one should have to make. When people in these circumstances need help, many of them turn to the HealthWell Foundation®. Founded in 2003, the HealthWell Foundation is an independent, non-profit organization that provides financial assistance to insured patients living with chronic and life-altering illnesses to help them afford their medical treatments.

Contact:

Phone: (800) 675-8416 | Website: <https://www.healthwellfoundation.org/fund/homocystinuria/>

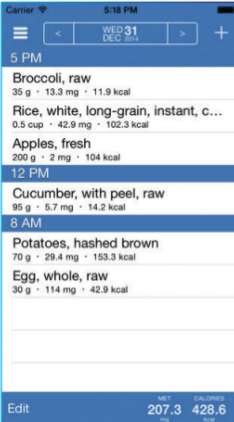
Diet Tracking Applications and Websites

HCU Network America does not endorse any diet tracking application or website.

AccuGo for HCU



Sponsored by
HCU
Network Australia



Time	Food Item	Protein (mg)	Methionine (mg)	Calories (kcal)
5 PM	Broccoli, raw	35 g	13.3 mg	11.9 kcal
	Rice, white, long-grain, instant, c...	0.5 cup	42.9 mg	102.3 kcal
	Apples, fresh	200 g	2 mg	104 kcal
12 PM	Cucumber, with peel, raw	95 g	5.7 mg	14.2 kcal
8 AM	Potatoes, hashed brown	70 g	29.4 mg	153.3 kcal
	Egg, whole, raw	30 g	114 mg	42.9 kcal

AccuGo for HCU was designed to ease the day to day management of the low protein diet by providing a quick and easy way to estimate and track protein or methionine in food eaten. It provides easy access to the protein and methionine content of foods including preloaded data, your own personal data and a community database of shared food.

Features

Daily and Historical Overview

- Provides an overview of the days running total of protein or methionine at a glance
- Navigate to the past to see historical entries
- Historical graph shows totals over the past month
- Navigate to the future to plan menus in advance

Search

- Search the database for the protein and methionine content of food including:
- Preloaded and identifiable data from both NUTTAB and USDA food lists
 - Your own personal food list
 - A Community Food List of shared food items within the community

Calculate

Provides automatic real time calculation of related values by entering your own quantities either in weight, protein, methionine or calories.

Food Lists

- Add your own foods and values to the database
- A Community Food List provides shared items you can add to your personal food list, or share yours with the community

Email Food Records

- Email daily food records by date range, includes summary of average over a specified period of time

Other features

- Track Methionine or Protein by changing mode in Options
- Online synchronization of food records between multiple devices
- Track up to four different individuals
- Continues to work offline, anywhere regardless of wifi or cellular availability
- Groups foods eaten together by selectable time periods (15mins, 1 hour, etc)
- Copy/move/delete entries with ease
- Built in calendar for easy navigation

Feedback

- Provide feedback so developers know where they are succeeding and where they can improve

Disclaimer
While all care has been taken in the preparation of the preloaded food items, no responsibility is accepted by the author(s) or HCU (Homocystinuria) Network Australia Inc., its staff or volunteers, for any errors, omissions or inaccuracies that may appear. No responsibility can be accepted by the author(s) or HCU (Homocystinuria) Network Australia Inc. for any known or unknown consequences that may result from reliance on any information provided on the Smartphone App. The Community Food List is a Food List that is contributed to by the Community and is not preloaded. We encourage you to check the accuracy when sharing a food with the community and when using the foods that appear on the community list.



Metabolic Diet App for HCY (homocystinuria, also known as HCU) is a website designed for patients with HCY. The website was designed with protein and methionine data provided by the Genetic Metabolic Dietitians International (GMDI) group to ensure accuracy and accessibility. In addition to the accurate data which can be used to track your daily nutrient intake, you can add your own foods and recipes, and export your diet records.

To find out more about Metabolic Diet App for HCY or to create an account, please visit www.metabolicdietapp.org/HCY.html

HOW MUCH PHE

HowMuchPhe.org is a comprehensive diet-management system for patients with PKU with phe/pro/exchange and caloric values for more than 7,000 foods. The How Much Phe website allows you to search their data base, find related values, calculate your own recipes and create a daily food log. In addition, your subscription comes with responsive customer service. While this website is designed for patients with PKU, it's a valuable resource because of the partnerships they have developed with many food companies to obtain unrounded protein content, which otherwise is hard to find.

PKU Diet Management at your fingertips

HowMuchPhe.org

A comprehensive diet-tracking system with phe/pro/exchange and calorie values for more than 7,000 foods

EASY FOOD SEARCH & DIET TRACKING

- Search and find phe values for 6500+ foods. Add common foods to an easy-access favorites list and add foods not yet on the list.
- Calculate the phe/pro/exch content of foods based on their units (e.g. 10 pretzels or weight), or find out how much you can eat of a certain food by entering the amount of phe/pro/exch you have left to consume
- Easily record your intake, blood levels and other diagnostics. Track, phe, pro or exchanges. Includes support for the simplified diet.

ADDITIONAL FEATURES

- Enter your own recipes & automatically calculate phe/pro/exchange & calories per serving
- Export data by email to share with your clinic
- Picture it: Get an easy visual of your levels and intake with the graphing and reporting tools
- Always ready: Available on any internet-connected device. All accounts are synced in real time.
- Multiple profiles for multiple PKUers in a family. Separate logins for parents and caregivers.

COST AND DEMOS

- \$45 annual fee supports food data maintenance, Amino Acid Analysis, and National PKU News programs
- Free demo accounts available (full functionality, limited number of foods)
- Credit card required to sign up. If you don't have a credit card or can't afford the annual fee, ask your clinic about sponsoring your subscription or go to howmuchphe.org/scholarships

HOW MUCH PHE  PKU News



QUOTES FROM HOWMUCHPHE.ORG USERS

"I hope you know you have changed so many people's lives. I have a 7 month old with classical PKU. We're so lucky to have kids in this generation where there are so many foods available but **we are even luckier to have HowMuchPhe.**"

BABIES

"HMP is an amazing database of information for those with metabolic conditions (my son has Tyrosinemia). **The ease of use to look up items in a supermarket or at a party makes keeping track of phe effortless.** We have trained my son's grandparents and aunts on how to use it so when they care for him, they can look up and enter food as well."

TODDLERS

"The time has come to let my 9-year-old go away and manage her own diet for a weekend. **This would never happen without How Much Phe.**"

PRE-TEENS

"How Much Phe is a game changer in the PKU community. The ability to track phe at your fingertips, to have my daughter enter in something that she had at lunch so that I can plan dinner accordingly makes this very very complicated diet that little bit simpler, that little but more manageable. **It is an immense help in the transition for my teenager toward independent dietary management and for that I am forever grateful!!**"

TEENAGERS

"Although I've been on diet my whole life, my levels have always been high. I've been on a mission to get my levels low for the first time with food documentation and measurement, more blood draws and increased formula. **HowMuchPhe has changed my life!**"

ADULTS

"HMP is an amazing tool! It is freeing to not have to lug around a large paper book. **It makes it easier to eat out and to travel.** And it is reassuring to know that the entries are being updated often."

Doctor's information

Geneticist:		Clinic Affiliation:	
Address:		City, State, Zip code:	
Phone:		Email:	
Dietitian:		Clinic Affiliation:	
Address:		City, State, Zip code:	
Phone:		Email:	
Primary Care Physician:		Clinic Affiliation:	
Address:		City, State, Zip code:	
Phone:		Email:	

