

Returning to Diet:

MANAGEMENT TIPS & POINTERS
FROM ADULTS WITH PKU



Based on Input from Adults with PKU of all Ages and Situations



PKU Adult Program

www.npkua.org

Made Possible by a Generous Donation from
Art and Catherine Higgins, parents of two late diagnosed PKU children
Compiled by Annaliese Martinec, Graphics By Kinnon Martin

Congratulations on taking this important step towards managing your PKU!

By returning to diet or tightening your compliance with the diet, you have chosen a path that will lead you to a more healthy and productive lifestyle. You may have felt like you were the only one with PKU on the planet growing up, but **you are not alone**. One person in every 15,000 Americans has Phenylketonuria (PKU). Chances are, there is someone near you going through similar challenges.

The National PKU Alliance Adult Program is here to help you. This booklet was **created by** adults with PKU, **for** adults with PKU, and, **especially for** adults like you who are trying to return to the PKU dietary management.

We have designed this booklet as a simple reminder of **how** and **why** to follow a PKU treatment plan. You will find short, simple notes from other adults with PKU, both **diet for lifers** and **returning to dieters**, that can help you with your return to diet.

There Is A Plan

The first thing to know is that there is a management plan, and that it works.

There was a time when doctors believed it was ok to take patients off the diet. Research now shows that a treatment plan should be continued for life. Some people may recall that they “had PKU when they were a kid,” but PKU never goes away.

While treatment methods have changed over the years, physicians currently recommend a lifestyle that involves a diet of foods that are low in phenylalanine (PHE), a medical food (“formula”) that provides essential amino acids without PHE, and regular contact with a dietitian.

In some cases medications can be used, but the majority of the on-diet PKU community follows some variation of the low protein diet and medical food treatment.

Reminder: What Is PKU?



Phenylketonuria (PKU) is a genetic metabolic disorder in which enzymes in your liver do not break down the amino acid Phenylalanine (PHE) properly. PHE is a part of protein, which is the long lasting energy in food.

Words To Know:

Enzymes help the chemical reactions that break down food.

Phenylalanine (PHE) is one of the amino acids that makes up protein.

Protein is building material for your body; a source of energy found in foods.

How Does This Affect Me?

If your enzymes can't break down PHE completely, it collects in your blood and travels to your brain. This built up, unbroken PHE can cause harm to your brain that shows in a number of ways. Most common: depression, anxiety, headaches, and memory problems. Different people have different levels of enzyme activity, so different people experience different effects.

What If I Don't Feel Different?

Many people do not notice the effects until someone talks to them about the different signs and symptoms of not following the treatment plan or until their PHE levels come down and they can feel the difference themselves. When looking back at their time off diet, some people describe it as being in a fog.

Following a diet plan that involves foods that are low in protein helps your body manage the amount of PHE it has to break down. The formula is used to give you the rest of the parts of protein that you don't get from a low PHE diet.

***** VERY Important Note 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 to the health of the baby.

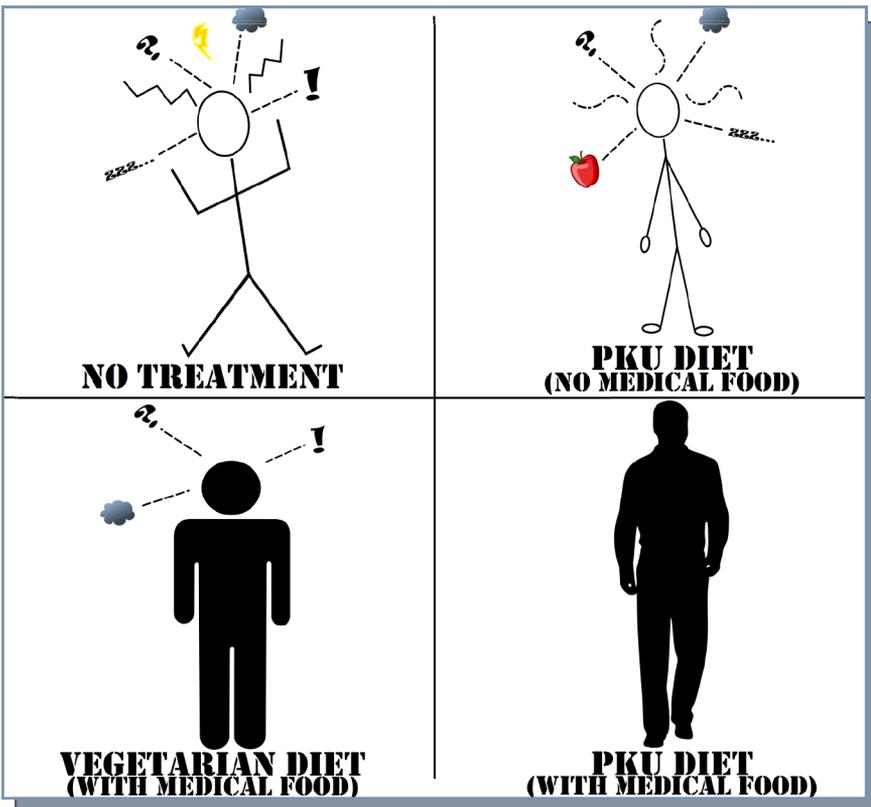
Symptom Checklist



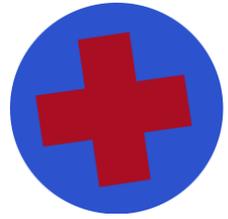
Do any of these sound familiar?

- Short term memory loss
- Trouble Focusing
- Headaches
- Depression
- Anxiety
- Eczema (skin rash)
- Hyperactivity
- Easily confused
- Paranoia
- Crying spells
- Difficulty with self control
- Difficulty with everyday tasks
- Lack of willpower
- Losing words when speaking
- Feelings of helplessness
- Fear of crowded places
- Constant sleepiness, lack of energy

With treatment you can overcome many of these symptoms over time.



Three Steps To Better PKU Management, Step 1: Clinic



When first returning to diet, it is important to visit a metabolic center that specializes in PKU because they will help you set and then reach your PKU goals. They can monitor the amount of PHE in your blood to find your PHE tolerance, help you find the best medical food for your needs, and provide guidance.

Your Phenylalanine tolerance is the amount of PHE that your body can handle each day.

1. Keep a diet record of everything you eat for three days. Look at the example of a breakfast below. **Don't be afraid** to tell your clinic what you're really eating. They are not there to judge you and will be able to help you better if they know where you're really starting from.
2. Have your blood drawn on the fourth day and sent in to a lab that can test for blood PHE level. Your clinic will help you arrange this.
3. The results will be sent to your clinic, where it will be recorded and shared with you by phone or email.

You may need to repeat this process a few times so your dietician and physician can provide you with an accurate PHE tolerance. Most clinics recommend that you have your PHE levels checked at least once a month so you can be sure everything is under control and to monitor how your levels respond to changes in your diet.

Date/Meal Quantity PHE Protein Food Description

Tuesday

Breakfast

2 scoops	0	--	PKU formula mixed in water
1 cup	6	0.2	coffee
1 tbs	12	0.2	non dairy liquid coffee creamer
1	5	0	apple
3	3	0	3-inch wide Mixquick Pancakes
2 tbs	0	0	syrup

26.0	0.4	Daily Totals
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Step 2: Make Friends With Your Formula



You Want Me To Drink 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 PKU it just happens to come in a different form.

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 PHE diet but not taking formula can leave you with severe nutritional deficiencies and feeling weak.

How Do I Get It?

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

Here's A Bit Of History:

The first PKU formula was introduced in 1951: Lofenalac. It was designed to supplement the protein needed during childhood, but many doctors had their PKU patients stop the treatment at adolescence. Fast forward through a lot of research to around 1980, when doctors began prescribing the PKU diet and medical food as a lifelong treatment. Medical food options were few at best, and were definitely not created for their taste. Thankfully the foods and formulas have improved greatly with time and effort, thanks to the hard work and dedication of the PKU professional and vendor community.

Step 2, Continued: Formula Tips And Tricks



I Tried This When I Was A Kid. 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, and cheers to a successful PKU diet!

Types Of Medical Foods

Formula has come a long way. For a long time our only choice was powder in a can but now we have better powder in a can, powder in single serving packets, pills and tablets, bars, and even ready-to-drink pouches and juice boxes! It comes in many flavors, too: lemon, orange, tropical, berry, vanilla, and even chocolate. Lots of people also choose unflavored formula and add their own flavoring.

Suggested Flavorings

Kool aid with sugar or Splenda, Tang, lemonade mix, Mio liquid water enhancer, iced tea, iced coffee, measured amounts of rice milk, or juice. There are a number of flavorings on the market now as well, just be sure to watch out for *aspartame!

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

*Watch out for the ingredient Aspartame. It should be avoided by people with PKU because it is made from Phenylalanine.

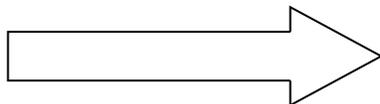
white	sugar	Sugar	safe
pink	saccharin	Sweet’N Low	safe
yellow	sucralose	Splenda	safe
blue	aspartame	Equal	NOT SAFE

When facing the colored sugar packets in restaurants, remember the blue ones are made with aspartame. The others are generally safe.

Step 3: What To Eat



People with PKU must avoid foods that are high in protein like meat, fish, poultry, dairy, soy, legumes (dried beans) or nuts. Some fruits and vegetables are higher in protein than others. PHE is in almost everything except sugar, salt, oil, and water. Thankfully, many fruits and vegetables are naturally low in PHE.



Check out the stoplight on the next page to get an idea which vegetables are within your PHE tolerance. Always be careful not to eat too much.

Foods in green have **less than 20mg PHE** in a 1/2 cup serving. These are great foods to fill up on.

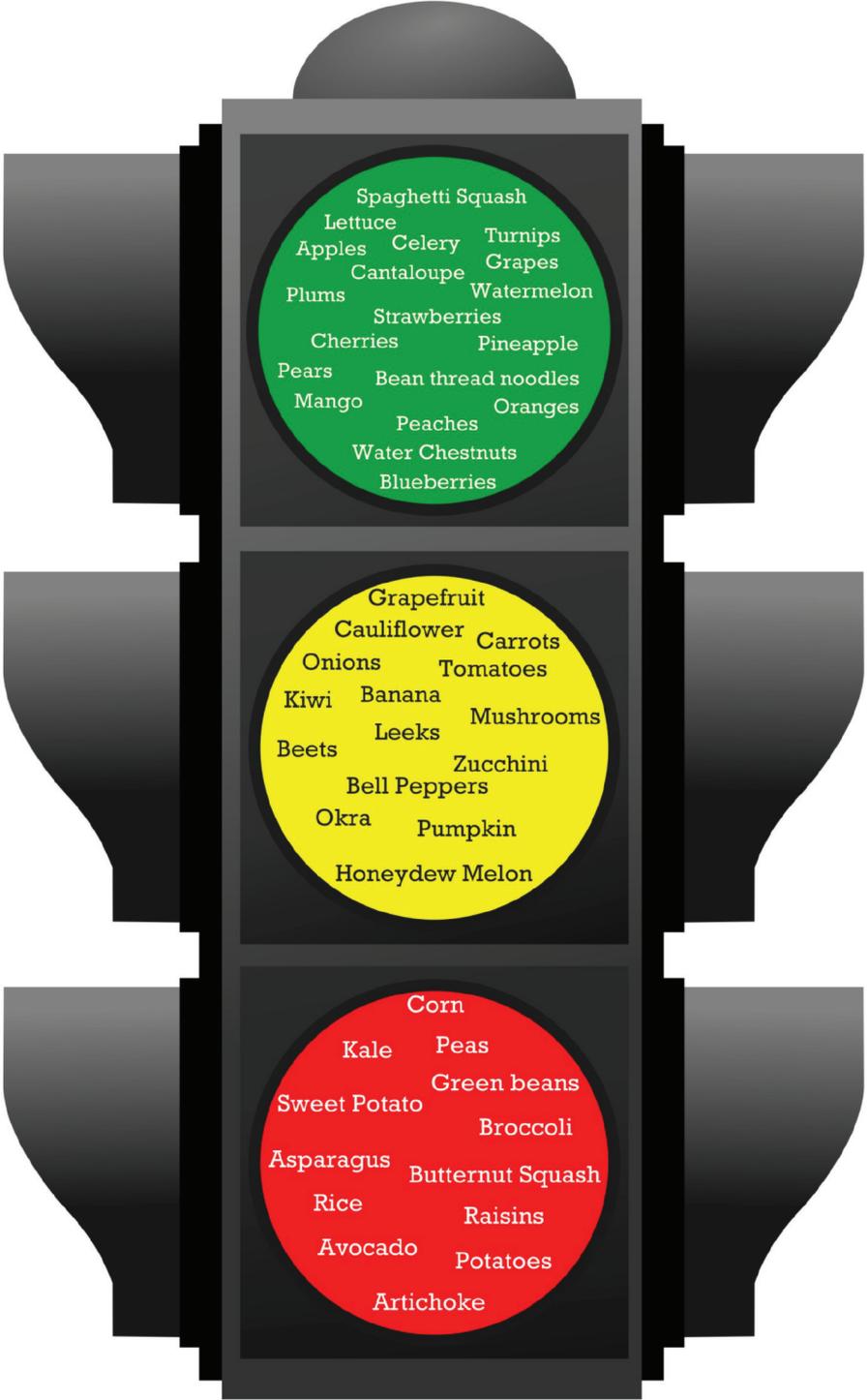
Foods in yellow have **21-50g PHE** per 1/2 cup serving. These can be eaten in moderation.

Foods in red should be eaten sparingly. They have **51-115mg PHE** per 1/2 cup serving.

Note: Some foods have very different PHE counts when they're cooked, from when they're raw. Different brands can also make a difference. Be sure to check a PHE list to get an accurate count. Your PKU clinic can tell you how to get a PHE list.

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 12 for a list of suppliers. Here we'll call them "**low pro foods**". You can find bread, 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 are more expensive, but provide much more variety. Some states and insurance companies offer help paying for low protein foods.



Spaghetti Squash
Lettuce
Apples Celery Turnips
Cantaloupe Grapes
Plums Watermelon
Strawberries
Cherries Pineapple
Pears Bean thread noodles
Mango Oranges
Peaches
Water Chestnuts
Blueberries

Grapefruit
Cauliflower Carrots
Onions Tomatoes
Kiwi Banana Mushrooms
Leeks
Beets Zucchini
Bell Peppers
Okra Pumpkin
Honeydew Melon

Corn
Kale Peas
Green beans
Sweet Potato Broccoli
Asparagus Butternut Squash
Rice Raisins
Avocado Potatoes
Artichoke

How To Count PHE in Foods



Here we will go over one of the more simple ways of counting PHE.

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

One gram of protein contains around 50 milligrams (mg) of PHE. This is a very rough estimate that can vary by 25mg more or less, but it is better than nothing.

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 1/2 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)

4. Multiply the total protein by 50 to get an estimate of the amount of PHE in the serving.

Note: It is very important to remember that any food containing Aspartame in the ingredients has Phenylalanine. Many foods that contain aspartame (like gum or diet soda) say they contain 0g protein, so don't be fooled. Food companies are not required to list how much PHE is in their products.

To be sure exactly how much PHE is in a food, you'll need to check a PHE list. The most widely used list is the Low Protein Food List for PKU by Virginia E. Schuett. See page 12 for ordering information. This book lists thousands of foods and their common serving sizes and PHE counts. It is a very valuable resource for anyone following the PKU diet.

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

Making Good Food Choices



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

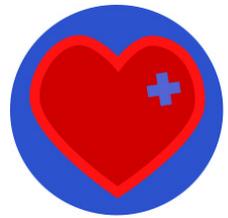
We even have more options in grocery stores now. You can often buy coconut milk yogurt, sliced cheese, and rice milk in normal grocery stores. Be sure to check the nutrition labels before you buy.

Small Changes

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

<u>Higher PHE</u>	<u>Lower PHE</u>
Bananas	Apples, grapes, berries
Creamy soups	Vegetable Soup
Ground beef	Chopped and seasoned mushrooms
Mashed Potatoes	Mashed cauliflower
Regular Yogurt or Soy yogurt	Coconut Milk Yogurt
Milk, half&half	Rice milk, non-dairy coffee creamer
Spaghetti	Bean Thread Noodles or Spaghetti Squash

Use Your Support System



Reach Out

Find and talk to others with PKU who are your age. 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 "PKU" in the search bar at the top of the page and saying hello to people who show up or joining some of the many PKU groups listed. Many states and regions have official support groups. Your clinic may be able to help you find them. Ask your clinic to put you in touch with other people with PKU and keep you informed about upcoming events. The National PKU Alliance has a list of support groups on their website.

Create Support

Support is so important. Many of the effects of PKU make starting your diet management really tough. Your family and close friends, peers or co-workers can provide a source of support. You don't have 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.

RESOURCES

INFORMATION & ASSISTANCE

National PKU Alliance
www.npkua.org
(715) 437-0477

National Organization for Rare Diseases
www.rarediseases.org
1-800-999-6673

Patient Power
www.patientpower.info
1-877-232-5445

PKU News
www.pkunews.org
(206) 525-8140

PKU FORMULA, FOOD & DRUG COMPANIES

Abbott Nutrition
www.abbottnutrition.com
1-800-227-5767

Applied Nutrition
www.medicalfood.com
1-800-605-0410

BioMarin
www.bmrn.com
www.PKU.com
1-866-906-6100

Cambrooke Foods
www.cambrookefoods.com
1-866-4-LOW PRO

Dietary Specialties
www.dietspec.com
1-888-640-2800

Ener-G Foods
www.ener-g.com
1-800-331-5222

Mead Johnson Nutrition
www.mjn.com
(812) 429-5000

Med-Diet
www.med-diet.com
1-800-MED-DIET

Nutricia
www.myspecialdiet.com
1-800-365-7354

PKU Perspectives
www.pkuperspectives.com
1-866-PKU-FOOD

Taste Connections
www.tasteconnections.com
(310) 371-8861

VitaFlo
www.VitaFloUSA.com
1-888-VITAFLO



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***The NPKUA works to improve the lives of individuals
with phenylketonuria and pursue a cure.***

The PKU Adult Program is a new initiative of the National PKU Alliance (NPKUA) that provides information and support for adults living with PKU. Adults with PKU face many challenges. The program provides a one-stop resource for adults seeking to improve their quality of life. This is the only program composed of a national network of adults with PKU, their families, clinicians and industry leaders to address the variety of needs of adults with PKU.