YOUR GUIDE

TO GETTING BACK ON THE PKU DIET

NUTRICIA DIET FOR LIFE™

IT'S NEVER TOO LATE

NUTRICIA

CONTENTS

David's story	1
PKU Q & A	2
Know your PKU facts	3
It's just not true...	4
Katie's story	5
PKU - Lets refresh	6
Nutricia's top 10 PKU formula for adults	16
Tips for making PKU formula part of your life	18
Taking the next step as easy as 1-2-3	20

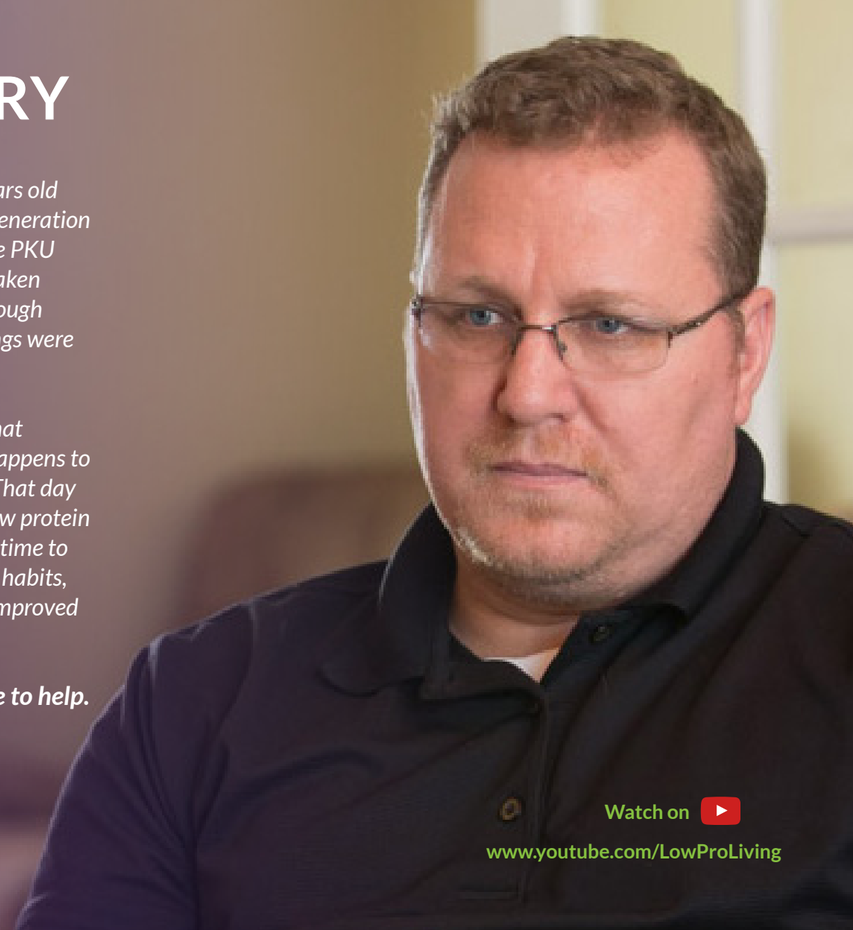
NUTRICIA DIET FOR LIFE™
IT'S NEVER TOO LATE

DAVID'S STORY

"My name is David Harris. I am 40 years old and I have classical PKU. I am a first generation PKU'er which means I was born before PKU was known to be Diet-for-Life. I was taken off-diet at the age of 6. I struggled through school and always wondered why things were so difficult for me."

"Then it happened. I saw a TV show that said PKU was diet-for-life and what happens to your body when you are not on diet. That day I knew I had to get myself back on a low protein diet and drink a PKU formula. It took time to re-learn the diet and adjust my eating habits, but when I went back on diet my life improved and now feel a lot better."

You can do it – and Nutricia is here to help.

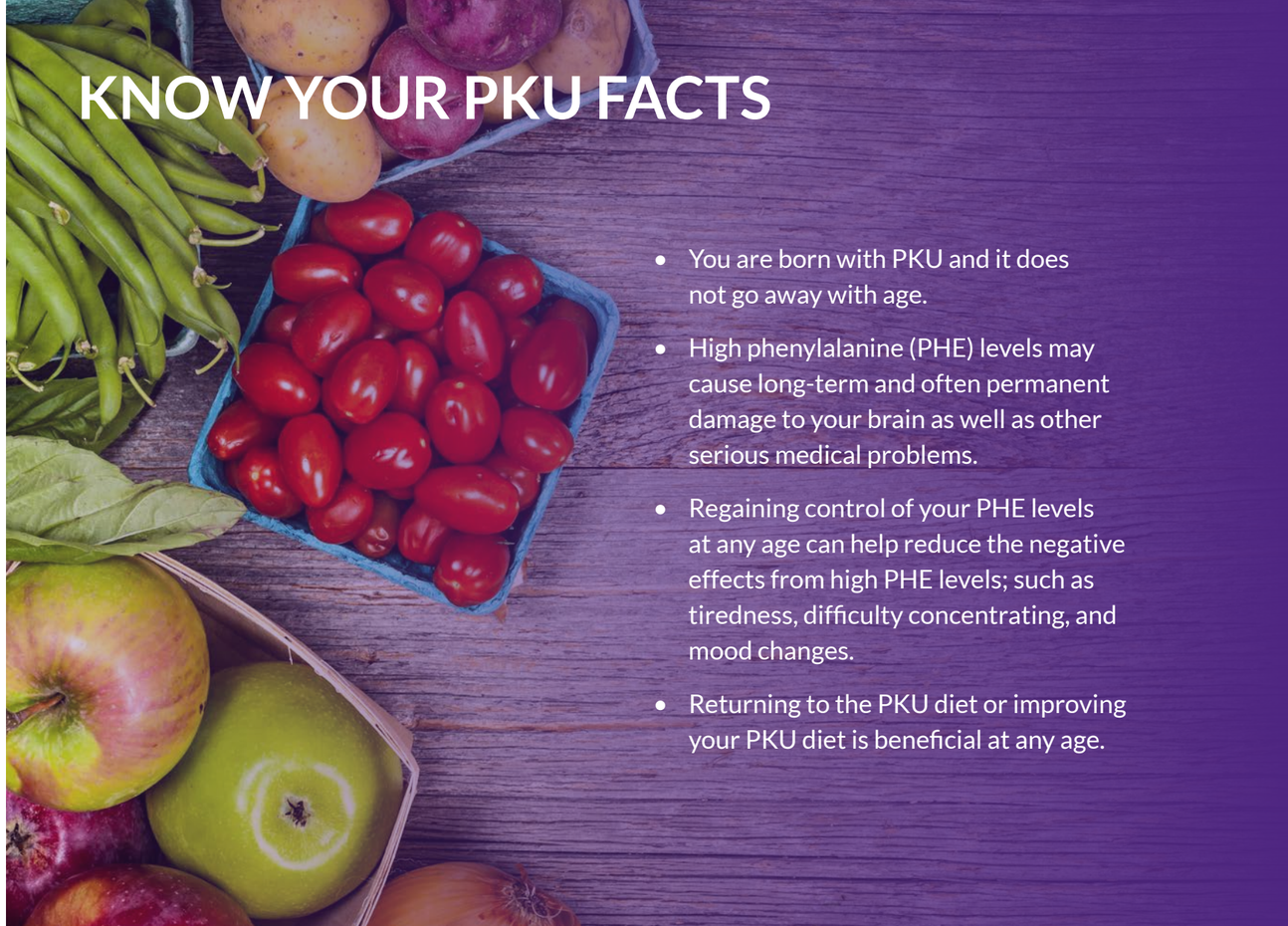


PKU Q&A

Please answer the following questions:

YES | NO	Did you follow a strict low protein diet and drink a PKU formula when you were younger?
YES | NO	Have you ever felt like there are limited choices in PKU formula?
YES | NO	Do you ever feel tired, foggy or irritable and wonder why or if it could be related to PKU?
YES | NO	Are you uncertain of how to obtain PKU formula and/or low protein foods?
YES | NO	Have you lost contact with the metabolic clinic you attended as a child?
YES | NO	Do you just avoid meat and other high protein food, but don't drink a PKU formula?

IF YOU ANSWERED YES TO 1 OR MORE OF THESE QUESTIONS, **THEN THIS BOOKLET WAS WRITTEN FOR YOU.**



KNOW YOUR PKU FACTS

- You are born with PKU and it does not go away with age.
- High phenylalanine (PHE) levels may cause long-term and often permanent damage to your brain as well as other serious medical problems.
- Regaining control of your PHE levels at any age can help reduce the negative effects from high PHE levels; such as tiredness, difficulty concentrating, and mood changes.
- Returning to the PKU diet or improving your PKU diet is beneficial at any age.

IT'S JUST NOT TRUE...

Just eating low protein foods without a PKU formula is an acceptable way to manage PKU.

FALSE. The truth is, you need a PHE-free protein source in your diet. PKU formula is a special modified protein source made specifically for you.

All PKU formulas and low protein foods taste bad.

FALSE. The truth is, PKU formulas have changed a lot in recent years. PKU formula options come in a variety of flavors, forms and styles. You can have a ready-to-drink formula that you can take on the go, or powders made with adults in mind that mix with a very small amount of water so you have less to drink. See page 11 to view PKU formula options.

I have to drink a lot of formula to get the full amount of daily protein I need.

FALSE. The truth is, many PKU formula options provide the daily protein you need in a more concentrated form (page 11).

I'm too old to go back to drinking PKU formula.

FALSE. The truth is, it's never too late to return to diet. Regaining control of your PKU diet at any age can help reduce the negative effects of PKU; such as tiredness, difficulty concentrating, and mood changes.

KATIE'S STORY

"When I was a teenager, I just wanted to be like everyone else. It started with pouring my formula in the sink. Then before I knew it, I was off diet."

"Since I could not immediately 'feel' my PHE levels going up, I didn't realize the changes that were happening. My grades got worse, my attitude and mood got worse. I was always tired and felt foggy."

"When I went back on diet – it was like the fog had lifted."

Katie, 31 - PKU

Watch on

www.youtube.com/LowProLiving

PKU – LET'S REFRESH

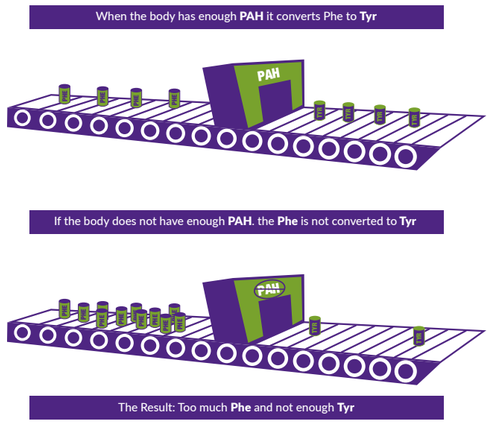
What is PKU?

Phenylketonuria (PKU) is a genetic disorder that a person inherits from both parents. PKU occurs due to a deficiency of the enzyme phenylalanine hydroxylase (PAH). This enzyme deficiency interferes with a person's ability to metabolize or process the amino acid phenylalanine (PHE) into tyrosine (TYR), another amino acid.

Amino acids, like PHE, are the building blocks of protein found in food and the human body. Consuming too much PHE from food can lead to a toxic build-up of PHE in the blood and brain of a person with PKU. Without management, this build-up of PHE will cause irreversible brain damage. Presently, there is no cure for PKU, but it can be managed effectively with proper diet and special nutritional formulas designed for persons with PKU.

Hyperphenylalaninemia

The Body is Similar to a Factory

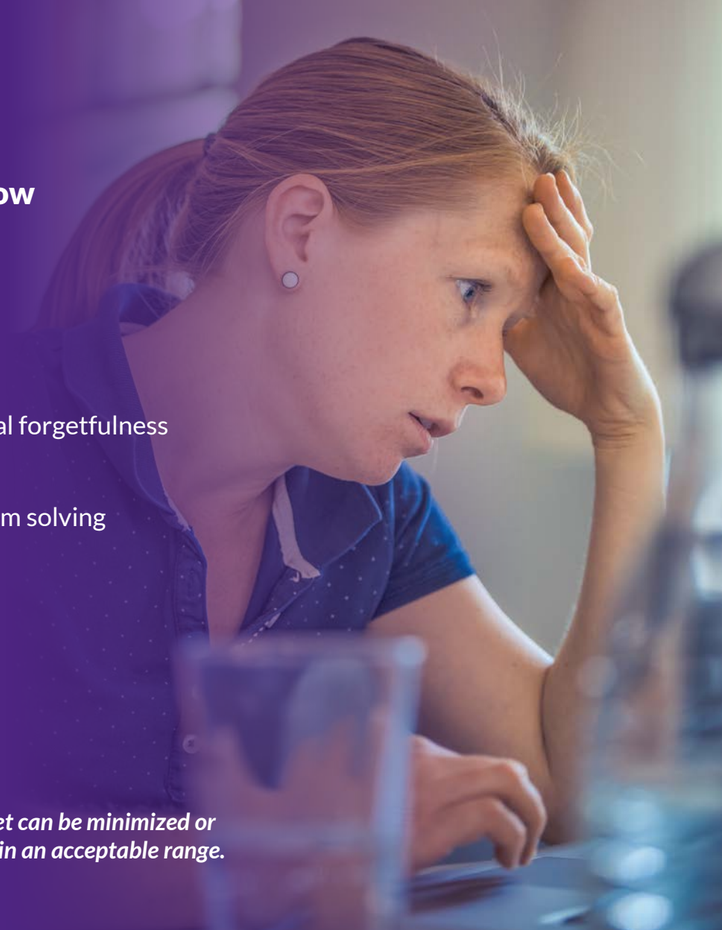


PAH: Phenylalanine Hydroxylase
Phe: Phenylalanine
Tyr: Tyrosine

Bernstein, L, Freehauf, C: Eat Right Stay Bright: Guide for Hyperphenylalaninemia. 1997-2001.

PHE build-up (high levels) and/or low levels of TYR can cause:

- Difficulty concentrating
- Feeling tired, moody or in a fog
- Tremors in hands, fingers, feet or legs
- Poor memory, short-term recall or general forgetfulness
- Trouble focusing at work or school
- Difficulty following directions and problem solving
- Anxiety, phobias or depression



Many of the problems associated with being off diet can be minimized or may go away once your blood PHE levels are back in an acceptable range.

What is the PKU diet?

The PKU diet is a diet that is low in natural protein to make sure you get just the right amount of PHE you need. Not too much to cause high blood and brain PHE levels; but also not too little as some PHE is necessary for normal body function.

All foods (other than pure fat, oil and sugar) contain some protein and PHE. Meat, chicken, fish and dairy are examples of foods high in protein and PHE. But other common foods such as bread, pasta, rice, potato, corn and peas also contain a fair amount of protein and PHE. Learning what foods are high in PHE and which are low will help with your long-term diet management.

However, just eating a diet low in natural protein would not cover the body's need for protein. So, to meet total protein needs, the PKU diet also includes PKU formula. PKU formula provides a source of safe protein with little or no PHE.

Why do I need the PKU diet?

The PKU diet is needed to help prevent a dangerous build-up of PHE. Check out the previous page again where we talked about signs and symptoms of high PHE and/or low TYR levels. Many teens and adults with PKU report feeling better when on the PKU diet.

MedicalFood.com

Shop at MedicalFood.com

NUTRICIA'S LOW PROTEIN FOODS

YOUR HOME FOR LOW PROTEIN PASTA, CEREAL, BAKING MIXES, AND MORE!

Free Shipping on all orders over $30!

Most fruits and vegetables are naturally low in protein and should be the biggest part of your diet. There are also special low protein foods that have been modified to be lower in protein and phenylalanine than their usual store-bought counterparts.

These special low protein foods, such as low protein pasta, bread, rice, muffins and baking mixes, are optional in the PKU diet, but they provide more variety and make the PKU diet more appealing and interesting.

What first steps can I take?

- Find a PKU formula that fits your lifestyle.
- Call a metabolic clinic. A dietitian will work with you to develop a personalized plan to help you get back on the PKU diet.
- Tell your family and friends. Talk to those closest to you about your dietary needs and goal to return to diet. This will help you develop a team of people to support you as you get back on diet or improve your current PKU diet.
- Meet other people with PKU, join a Facebook group, or attend a PKU event or conference.
- Fall in love with fruits and vegetables. Explore new items at your local market and look into new recipes online and in vegetable-focused cookbooks.

Don't have a metabolic clinic or remember your dietitian's name?

WE CAN HELP!

Email us at **DietforLife@Nutricia.com**

David, PKU

Sarah, PKU

Pedro, PKU

Katie, PKU

Where do I get a quality source of protein?

PKU formula is your main source of protein per day. Look at it as a special protein shake that contains a modified protein source specifically for those with PKU. The protein is typically a blend of amino acids or amino acids with GMP, that contain little to no phenylalanine. PKU formula is also supplemented with tyrosine, the amino acid that individuals with PKU can only make in small amounts. PKU formula must be consumed every day and used under medical supervision.

Your formula will make up the majority of your daily protein intake. Your healthcare professional will be able to tell you the amount that's right for you.

NUTRICIA NAVIGATOR

NUTRICIA IS HERE
TO LEND A HELPING HAND

ONE-ON-ONE SUPPORT TO OBTAIN FORMULA COVERAGE AT NO COST TO YOU!

As part of our Nutricia Connect program, we offer Nutricia Navigator, a free service available to help guide you through the world of formula coverage from start to finish.

- Contacting your health insurance plan to determine coverage
- Completing pre-determinations, prior authorizations and supporting medical necessity reviews
- Initiating joint calls between patients, healthcare teams and insurance companies to help guide you through difficult reimbursement processes
- Providing billing error support
- Assisting in appealing insurance denials for Nutricia products

To learn more visit
MedicalFood.com/Reimbursement or call **1-800-365-7354**

NUTRICIA CONNECT™
Formula Coverage • Education • Support

NUTRICIA'S TOP PKU FORMULA OPTIONS FOR ADULTS

NUTRICIA

SHAKES

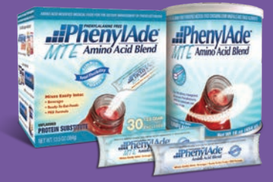
Contains GMP, made from a intact protein source

READY-TO-DRINK

Contains GMP, made from a intact protein source

LESS-TO-DRINK

Contains GMP, made from a whole protein source



ALTERNATIVE TO DRINKING FORMULA

ALTERNATIVE TO TRADITIONAL PKU DIET

TIPS FOR MAKING PKU FORMULA PART OF YOUR LIFE

Make a daily routine

- Your body does best when it receives nutrition in small amounts throughout the day. Aim to drink your PKU formula 3-4 times per day.
- Have trouble remembering? Set an alarm on your phone or use an alarm app to remind you to take your PKU formula.
- Consider a diet journal so that you can keep track of your intake throughout the day.

Plan ahead

- Mix your formula in advance. Most PKU formulas can be refrigerated up to 24 hours after mixing. Just remember to shake before you drink!
- Try a ready-to-drink PKU formula to save time.

Visit LowProLiving.com for recipes and mixing ideas.

Start the day off right

- Breakfast is an important meal of the day so be sure to have a serving of your PKU formula in the morning.
- Mix your PKU formula the night before if mornings tend to be a hectic time.
- You can blend in fresh or frozen berries to make a smoothie.

NUTRICIA DIET FOR LIFE™ IT'S NEVER TOO LATE

TIP: Use a clean, re-usable container that is not see through if you don't want others to know what you are drinking.

Don't leave home without it

- Make your PKU formula portable if your drink is not available in convenient single serving packages.
- Find small plastic containers or bottles that can hold 1-2 servings of formula that you can take with you.
- Pre-measure the powder and add water or a flavored drink when you are ready to have your PKU drink; or consider a product in pre-packed pouches.

Have a back-up plan

- Keep backup PKU formula in a convenient place to use as needed in case you forget or leave home without it.

Examples...

- In your bag or purse
- In your desk at work
- At your friend's house
- In your personal locker
- At the home of your parents or significant other

Be Creative

- Make a smoothie by adding ice cubes or frozen fruit to your formula in a blender.
- Freeze your PKU formula for 1-2 hours and eat it with a spoon.
- Mix formula into a cold, store-bought beverage while on the go.

Don't Run Out

- Keep track of how much PKU formula you have at home so you can pick up or order more in advance.
- If possible, make a standing order with your pharmacy or PKU formula provider, so you get a shipment every month and never run out.
- Leave yourself at least a 2-week supply of formula; just in case there is a delay in your shipment.

TAKING THE NEXT STEP AS EASY AS 1-2-3

1 Call Nutricia and speak to a product specialist

NUTRICIA CAN HELP YOU:

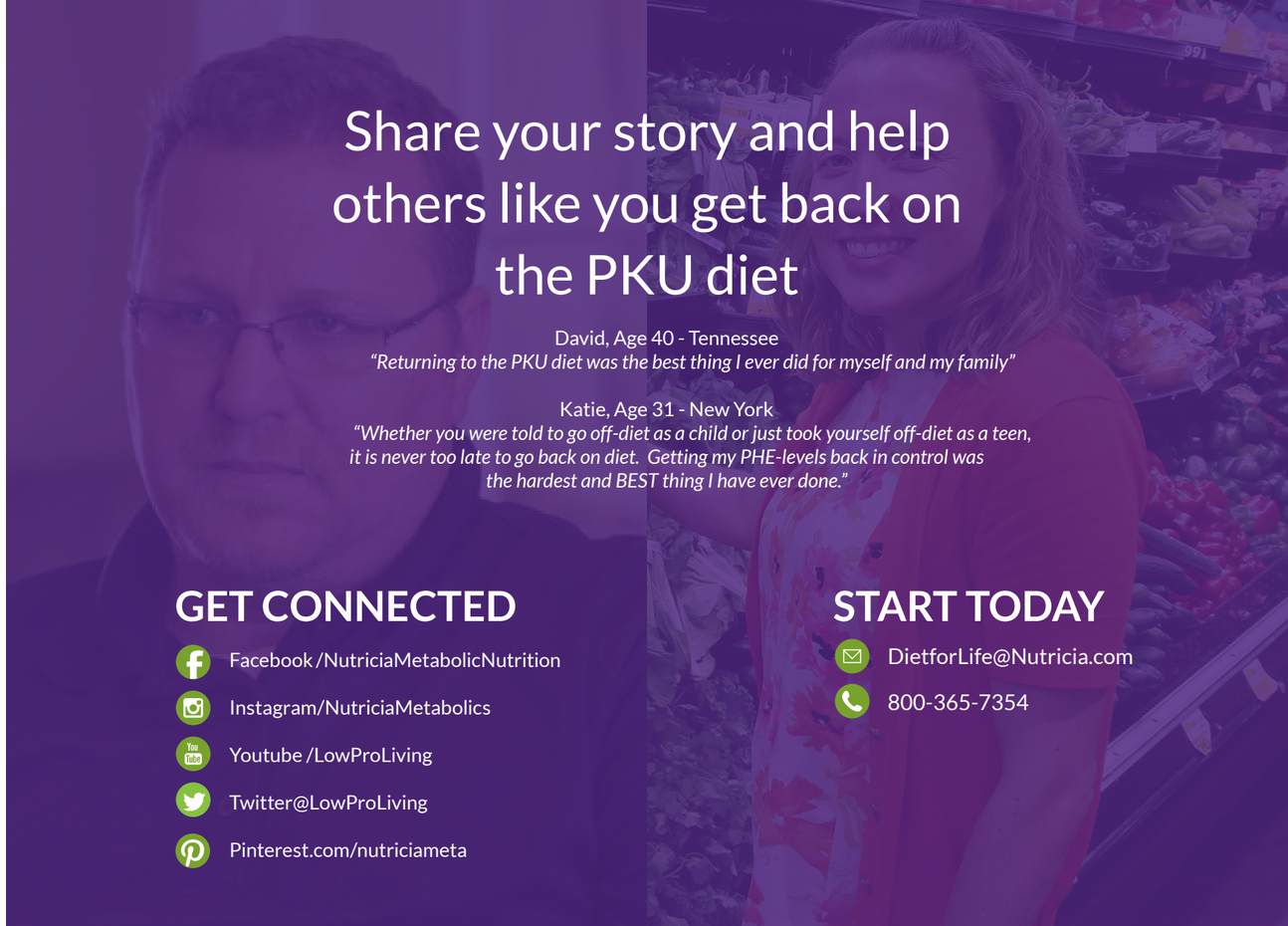
- Learn more about returning to diet and advances in PKU products
- Find a PKU formula to match your personal needs
- Connect or re-connect with a metabolic healthcare professional
- Determine if your insurance will cover your PKU formula
- Locate a PKU formula supplier once coverage is determined
- Discuss alternatives if you do not have medical insurance

2 Start making the PKU diet part of your daily routine

- Set a menu for low-protein meals
- Hit the market and stock up on fresh fruits and vegetables
- Make a plan and set specific times for drinking your formula

3 Stay in tune with how you are feeling

- Talk openly with friends and family
- Ask if they see improvements in you and/or your behavior
- Set small goals, like staying on diet for a full day, then a week, then a month
- Celebrate you and your success



Share your story and help others like you get back on the PKU diet

David, Age 40 - Tennessee
"Returning to the PKU diet was the best thing I ever did for myself and my family"

Katie, Age 31 - New York
"Whether you were told to go off-diet as a child or just took yourself off-diet as a teen, it is never too late to go back on diet. Getting my PHE-levels back in control was the hardest and BEST thing I have ever done."

GET CONNECTED

- Facebook /NutriciaMetabolicNutrition
- Instagram/NutriciaMetabolics
- Youtube /LowProLiving
- Twitter@LowProLiving
- Pinterest.com/nutriciameta

START TODAY

- DietforLife@Nutricia.com
- 800-365-7354

This book is dedicated to Dr. Richard Koch, a PKU pioneer who helped save countless lives and helped bring hundreds of adults back to the PKU diet.

Prior to the 1980's it was believed that children as young as 5 years of age could stop taking their PKU formula and continue life on a liberalized diet without any repercussions. However, over time, with having the results from numerous clinical studies, it was determined that a low-phenylalanine diet with PKU formula should be followed for life. These studies showed that those who discontinued their PKU formula and liberalized their diet had problems such as recurrent headaches, neurological abnormalities, hyperactivity, lethargy, phobias and depression.
As a result, it is now widely accepted that PKU is Diet-for-Life.

NUTRICIA
DIET FOR LIFE™
IT'S NEVER TOO LATE

All PKU formulas shown are medical foods for the dietary managment of proven Phenylketonuria (PKU) and must be used under medical supervision.

© 2019 Nutricia North America 03/20 ZPKUDLB

NUTRICIA