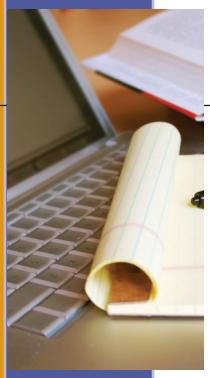

Chapter 15: Resources

Sample Letters for School and Other Activities

The following sample letters of medical necessity should be provided by a PKU team member. He or she can prepare a customized letter for you. Keep a letter of medical necessity with you when you are travelling to ensure you can easily respond to any questions asked at airport security or customs.

SAMPLE LETTER FOR INSURANCE COVERAGE OF FOODS MODIFIED TO BE LOW IN PROTEIN

(Date)

RE: (patient name)
D.O.B: (patient date of birth)

To Whom It May Concern:

We are writing a letter of medical necessity regarding the treatment of (patient first name & last name). (patient name) has been under the consultative care of the (clinic name). He/She has an inborn error of metabolism, a genetic disorder, known as phenylketonuria (PKU, ICD 9 270.1). We are writing to request that low protein modified food products be covered by his/her current medical insurance.

PKU is a lifelong problem that requires a phenylalanine-restricted diet including low protein modified food products and the prescription of medical foods/formulas by a licensed physician with the support of a registered dietitian in order to control the blood phenylalanine level. Low protein modified food products are defined as manufactured products that will deliver no more than one gram of protein per serving. Low protein modified food products supply needed additional calories (to help prevent catabolism, which in itself can cause phenylalanine levels to rise), without supplying additional phenylalanine containing protein. Use of low protein modified food products, especially when used consistently, greatly improves adherence to the treatment program.

PKU results from a deficiency of the enzyme responsible for metabolizing the amino acid phenylalanine. This results in the build-up of phenylalanine to toxic levels. An untreated child with PKU will suffer irreversible brain damage as well as severe and progressive neurological disorders. Normal growth and development are possible if an infant with PKU is treated appropriately. In adolescents and adults, neurological deterioration, phobias, difficulty in concentration and impulse control, and loss of IQ points can occur if treatment is not sustained.

Patients are treated with prescribed medical foods/formulas, as well as a phenylalanine-restricted diet which includes low protein modified food products. This diet excludes all foods high in protein (i.e. meat, poultry, fish, dairy, nuts and legumes) and markedly restricts all grains, including rice, breads, and pastas. Medical foods/formulas provide the primary protein constituent (80-85% of RDA protein) for the PKU dietary treatment regimen. Low protein modified food products other nutrients which includes additional calories to prevent catabolism which can cause a rise in phenylalanine levels. Use of these products is medically supervised by a physician and implemented by a registered dietitian specially trained in the nutrition management of inborn errors of metabolism. Nutrition therapy must also

provide a sufficient and balanced intake of other nutrients to avoid nutritional deficiencies. Nutrition therapy of PKU solely via protein restriction is not possible, because it will result in protein malnutrition, calorie deprivation, vitamin and mineral deficiency, failure-to-thrive, and potentially death.

The standard of care for PKU requires the use of the medical food/formulas and a phenylalanine-restricted diet which includes the use of low-protein modified food products, as well as routine nutrition follow-up with a specially trained registered dietitian. The two primary goals of treatment are:

1. To maintain the blood phenylalanine at a level that is not toxic, but still allows for normal growth and development.
2. To ensure that the individual's overall nutritional requirements are met, allowing for normal growth and development, and the avoidance of nutritional deficiencies.

The recommended treatment range of blood phenylalanine levels for individuals with PKU is between 2 and 6mg/dL (120 and 360µmol/L). There is good correlation of cognitive function and maintenance of blood phenylalanine levels in this treatment range. Elevated blood phenylalanine in patients has been associated with behavior and learning problems which can reverse when the blood levels return to the treatment range. Currently, indefinite continuation of dietary management is recommended to all patients with PKU. These recommendations are based on a growing body of evidence indicating there is a decline in average IQ and development of difficulties in school performance after diet discontinuation.

We appreciate your attention to this request for (patient's name)'s low protein modified food products to be covered by his/her current medical insurance. Please do not hesitate to contact us if you have any questions at (clinic contact info).

Sincerely,

(dietitian name), RD, LDN

(Physician name), M.D.
(physician credentials, clinic name)

cc: (parents name)

Resources

SAMPLE LETTER FOR INSURANCE COVERAGE OF MEDICAL FOODS

(Date)

RE: (patient name)
D.O.B: (patient date of birth)

To Whom It May Concern:

We are writing a letter of medical necessity regarding the treatment of (patient first name & last name). (patient name) has been under the consultative care of the (clinic name). He/She has an inborn error of metabolism, a genetic disorder, known as phenylketonuria (PKU, ICD 9 270.1). We are writing to request that medical food/formula be covered by his/her current medical insurance.

PKU is a lifelong problem that requires a phenylalanine-restricted diet and the prescription of special medical foods/formulas by a licensed physician with the support of a registered dietitian in order to control the blood phenylalanine level. The term medical food/formula as defined in section 5(b) of the Orphan Drug Act {21 U.S.C. 360ee (b) (3)} is a "food which is formulated to be consumed or administered internally under the supervision of a physician and which is intended for the specific dietary management of a disease or condition for which distinctive nutritional requirements, based on recognized scientific principles are established by medical evaluation."

PKU results from a deficiency of the enzyme responsible for metabolizing the amino acid phenylalanine. This results in the build-up of phenylalanine to toxic levels. An untreated child with PKU will suffer irreversible brain damage as well as severe and progressive neurological disorders. Normal growth and development are possible if an infant with PKU is treated appropriately. In adolescents and adults, neurological deterioration, phobias, difficulty in concentration and impulse control, and loss of IQ points can occur if treatment is not sustained.

Patients are treated with prescribed medical foods/formulas (in a variety of forms powder, capsule, liquid, bar etc.), special low-protein modified food products as well as a phenylalanine-restricted diet. This diet excludes all foods high in protein (i.e. meat, poultry, fish, dairy, nuts and legumes) and markedly restricts all grains, including rice, breads, and pastas. Currently, (patient name) is prescribed (name of medical formula) which is a medical formula used to manage PKU. Medical foods/formulas provide the primary protein constituent (80-85% of RDA protein) for the PKU dietary treatment regimen. Use of these products is medically supervised by a physician and implemented by a registered dietitian specially trained in the nutrition management of inborn errors of metabolism. Nutrition therapy must also provide a sufficient and balanced intake of other nutrients to avoid nutritional deficiencies. Nutrition therapy of PKU solely via protein restriction is not possible, because it will result in protein malnutrition, calorie deprivation, vitamin and mineral deficiency, failure-to-thrive, and potentially death.

The standard of care for PKU requires the use of the medical food/formulas and a phenylalanine-restricted diet, as well as routine nutrition follow-up with a specially trained registered dietitian. The two primary goals of treatment are:

Resources

1. To maintain the blood phenylalanine at a level that is not toxic, but still allows for normal growth and development.
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The recommended treatment range of blood phenylalanine levels for individuals with PKU is between 2 and 6mg/dL (120 and 360µmol/L). There is good correlation of cognitive function and maintenance of blood phenylalanine levels in this treatment range. Elevated blood phenylalanine in patients has been associated with behavior and learning problems which can reverse when the blood levels return to the treatment range. Currently, indefinite continuation of dietary management is recommended to all patients with PKU. These recommendations are based on a growing body of evidence indicating there is a decline in average IQ and development of difficulties in school performance after diet discontinuation.

We appreciate your attention to this request for (patient's name)'s medical formula, (name of medical formula) to be covered by his/her current medical insurance. Please do not hesitate to contact us if you have any questions at (clinic contact info).

Sincerely,

(dietitian name), RD, LDN

(Physician name), M.D.
(physician credentials, clinic name)

cc: (parents name)

Resources

SAMPLE LETTER FOR TRAVEL

Date: _____

RE: _____ traveling with medical formula and low protein foods

To Whom It May Concern:

My patient,_____, is a _____year old boy / girl with an inborn error of metabolism called phenylketonuria (PKU). The treatment for PKU is dietary management and restriction of dietary
phenylalanine (an amino acid) and daily consumption of a medical nutritional formula.

_____'s medical formula is a mixture of a formula called_____ For a PKU
patient, it is crucial for all this formula to be consumed daily with special low protein foods which are part of treatment for PKU.

During _____'s travels – including the flight - it is essential that he / she have access to the formula (in powder and liquid form) and specialty low protein foods. They will bring what they need with them on the plane, including the formula, and the rest will be in their checked bags. The following non-perishable formula and perishable/non-perishable food items will be brought with them during their travels:

- Name of medical formula: _____ (in powder and or liquid form)
- Low protein foods _____
- Gram scale to measure foods and formula

Please allow the family of _____to bring formula and low protein foods on the plane during their travels so that they may properly care for their child while on vacation.

_____'s medical care is coordinated by Dr._____ and the clinic staff at_____. If you have any questions

Sincerely,

_____

Dr.

Resources

Handouts

For any parent, their child's first time experiences like having dinner with friends or sleepovers can be stressful. For the parent of a child with PKU, it can be especially stressful. You may worry about whether your child will be able to eat anything, or if he or she will end up eating something that is inappropriate for a low Phe diet.

You may not be the only one. It can be stressful for a new daycare provider, teacher, parent of a friend or babysitter to know that a child in their care has a special diet.

An easy way to help put your mind at ease – and theirs – is to provide them with a handout of information. Consider giving them a copy of the "FAQs" as well, if that level of information is required, or, if your child will be with them for a meal, fill out the blank menu form that will help them prepare a simple meal that is nutritious and appropriate for your child with PKU.

Consider meeting with people personally; reassure them that you are there to provide support, especially for new daycares and schools.

Resources

PKU Information for Adult Friends of the Family

You will be in close contact with_____ who is a healthy child, but has a rare genetic disorder called phenylketonuria or PKU. This means that _____has special dietary requirements that are very important, but is otherwise very healthy and can do anything else that other children enjoy. It's important that you feel comfortable and confident that you can handle the diet, too, so here is some information you might find helpful.

Here are some key points that you may want to know:

- PKU is a genetic condition that is not contagious.
- Apart from needing a special diet, a person with PKU is healthy.
- People with PKU cannot break down an amino acid called phenylalanine (Phe), which is found in all foods containing protein.
- Phe can build up in the blood and damage the developing brain.
- Staying on a low protein diet keeps Phe levels in a safe range, allowing for normal development and a healthy life.
- Eating the wrong foods will not make a person with PKU sick right away, but will cause problems over the long-term. Having food that is not part of the diet should not be considered a "treat" as it will have implications for an individual with PKU.
- A person with PKU does not outgrow it and must stay on the diet for life.

Foods that are safe as snacks are_____ _____. It's important that we always know what_____has eaten, so please let us know if any snacks have been served, and how much. We will record this so we can make sure_____has all the nutrients needed every day. If ever _____will be over for dinner, we can send a PKU meal, or an easy low Phe menu for you to prepare. And please don't be afraid to ask any questions you may still have!

Resources

PKU Information for Friends of a Child with PKU

Hi! My name is _____. I have a disorder called phenylketonuria or PKU. This means that there are some things I can't eat – kind of like having allergies. I have a special drink that helps make sure I grow and have big muscles. You will see that I drink it a lot!

Some things you may want to know are:

- You can't catch PKU. People are born with PKU, like I was.
- Having PKU means I have to eat foods that keep me healthy.
- I can't eat some kinds of foods, like meat or cheese, or they will make me sick. I won't get sick right away, but if I eat these foods, my body and brain won't grow the way they are supposed to.
- I drink a special milk which is like a vitamin drink. It gives my body good things that come from foods I can't eat.
- Having food that I'm not allowed isn't a "treat", but there are treats that I can have – like_____!
- I will always have PKU, it won't go away, so I will always be on a special diet.
- I can run, jump, play and do anything any other kid can do!

Just so you know:

- My favorite treat is _____ .
- My favorite activity is_____ .
- My favorite toy is _____ .

In case you have any questions or want any more information, you can call:

_____at_____ .

Resources

PKU Information for Babysitters

As a babysitter for_____, you may wonder what phenylketonuria (PKU) is_____. is a healthy child, but has a rare disorder called phenylketonuria or PKU. This means that_____ is healthy, but has a special, very important diet. It's important that you understand this, so I'd like you to review some information that will help you while babysitting_____.

Here are some key points that you may want to know:

- PKU is not contagious.
- Apart from needing a special diet, a person with PKU is healthy. It's kind of like having allergies, but sometimes more serious.
- People with PKU cannot break down parts of protein – which is something found in meat, eggs, milk and a lot of other foods.
- Phe can build up in the blood and damage a person with PKU.
- Eating the wrong foods will not make a person with PKU sick right away, but will cause problems over the long-term. Having food that is not part of the diet should not be considered a "treat."
- A person with PKU does not outgrow it and must stay on the diet for life.

When we leave you with_____, we will leave out prepared meals and some foods that are safe as snacks such as_____. It's important that you do not give_____ any other food than what has been provided or approved at any time as it can make him or her sick.
It's also important that we always know what_____has eaten, and how much, so please let us know what has been eaten (or if something hasn't been eaten that we left out).

Please don't be afraid to ask any questions you may still have about_____.
It's important to us that you're comfortable and able to help us with_____'s special diet.

Resources

PKU Information for Daycare Providers and Teachers

As daycare provider for_____, you will be responsible for _____independently. This is an important responsibility, and I want to make sure you are comfortable in this role. This is some information to help you understand phenylketonuria (PKU) and how it affects my child.

_____is a healthy child. PKU is a rare genetic disorder. This means _____that has special dietary requirements that are very important, but is otherwise very healthy and can do anything else that other children enjoy.

Here are some key points that you may want to know:

- PKU is a genetic condition that is <u>not</u> contagious.
- Apart from needing a special diet, a person with PKU is healthy.
- People with PKU cannot break down an amino acid called phenylalanine (Phe), which is found in all foods containing protein.
- Phe can build up in the blood and damage the developing brain.
- Staying on a low protein diet keeps Phe levels in a safe range, allowing for normal development and a healthy life.

- Eating the wrong foods will not make a person with PKU sick right away, but will cause problems over the long-term. Having food that is not part of the diet should <u>not</u> be considered a "treat" as it will have implications for an individual with PKU.
- A person with PKU does not outgrow it and must stay on the diet for life.

We will work with you to help you learn the details about planning meals for PKU. We can also review the menus that you prepare, identify what _____can eat, and supplement it with what is required for a nutritional PKU diet.
Foods that are safe include_____. We can provide treats that are appropriate for _____so that when special events like birthdays happen, he or she will have a treat to enjoy as well, that's safe for him or her to consume.

It's important that we always know what_____ has eaten, so please let us know everything what_____has eaten, and how much. This

includes if he or she has not eaten some of the food that has been provided as we have to make up for it throughout the day. If any Phe-free snacks have been served, we need to know this, too. We will record this so we can make sure_____has all the nutrients needed every day.

Attached is a list of Frequently Asked Questions. Please don't hesitate to ask any additional questions you may have. We will work closely with you to assist with this transition in any way possible.

Sincerely,

Contact information: _____

FAQs for Adults[99]

Q. How common is phenylketonuria (PKU)?
A. Approximately one child in every 15,000 births has PKU[100]. All states have effective PKU newborn screening programs, so most children with PKU start a low protein diet within the first month of life. The diet is well-controlled to ensure that the children grow and develop normally and attend regular schools.

Q. Since PKU is inherited, do all the children in that family have PKU?
A. No, there is a 25 percent chance for other children to have PKU. For a child to have PKU, one parent must have PKU or each of the parents must be non-symptomatic carriers of the PKU gene meaning that they have one normal gene and one PKU gene. Two carrier parents have a 25 percent chance of having a baby who is free of the PKU gene, a 50 percent chance that the baby will be a carrier, and a 25 percent chance the baby will have PKU. Families with one child with PKU may have others that do not.

Q. How is the PKU diet planned?
A. The diet depends on how much Phe is allowed in the diet and how much of the medical formula the individual needs. Below is a typical low protein menu.

MEALS	FOOD ITEMS	PHENYLALANINE
Breakfast	3 CBF Mixquick pancakes	2
	2 tbsp Pancake syrup	None
	1 Apple	11
	Medical formula	None
Lunch	1 ½ cups (cooked) Loprofin Spaghetti	7
	½ cup Marinara Sauce	39
	1 cup Iceburg Lettuce	14
	2 tbsp Italian dressing	None
	Medical Formula	None
Dinner	1 PKU Perspectives Chicken-flavored patty	39
	1 CBF bun	6
	½ cup Fried potatoes	76
	2 tbsp Ketchup	18
	10 baby carrots	30
	1 Mandarin Orange	16
	Medical Formula	None
TOTALS		**258 mg(~5.2g protein)**

Q. How long will they be on this diet?
A. A person with PKU is put on a strict low Phe diet as soon as he or she is diagnosed with PKU. It is recommended that anyone with PKU remain on their strict diet for their entire life, as high Phe levels can cause health problems for a person with PKU at any age.

Q. Can a child grow with this strict diet?
A. The formula or drink contains most of the protein, vitamins and minerals needed for growth. The food eaten provides the rest of the nutrients needed, and there are "free foods" that do not contain Phe that a person with PKU can eat when he or she is hungry.

Q. Can a person with PKU have any treats?
A. Some treats a person with PKU can have include lollipops, popsicles (without ice cream) and other sugar candies.

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[99]Kaufman, M, Nardella, M. *A Teacher's Guide to PKU* Texas Department of Health Available At: http://www.ub.edu.ar/centros_de_estudio/ceegmd/documentos/TeachersGuide.pdf Accessed June 15, 2011
[100]National Institutes of Health. *Phenylketonuria: Screening and Management National Institutes of Health Consensus Development Statement* Available At: http://consensus.nih.gov/2000/2000 phenylketonuria113html.htm Accessed May 17, 2011

Q. Does a child with PKU look or act differently from other children?
A. No, a child with PKU is just like other children in your classroom except that he/she has a special diet.

Q. How can I explain the PKU diet to children?
A. Young children can understand that since cars with different engines use different fuel (gas, diesel, etc.), some children have bodies that work in different ways than others and they need different food. Older children can understand the similar concept of a "food allergy." Don't hide the fact that the PKU child's lunch is different if asked, but no long explanation is needed. Ask the child's family for suggestions on how best to answer this question. You may also want to review our list of questions that children may encounter to prepare yourself for young, curious minds.

Q. If a child with PKU eats a high protein food, will he or she feel sick?
A. If a PKU child does eat a high protein food, he/she will probably not feel sick or different in any way. It is the long term, that elevated blood Phe level interferes with mental development. People with PKU may report feeling irritable and may have difficulty paying attention if they are not following the det completely. The changes may not be seen for several months to a year or more.

Q. What is the connection between aspartame and PKU?
A. Aspartame is used as a sweetening sugar substitute. When aspartame is broken down in the body, over half of it is Phe. Since individuals with PKU limit Phe in their diet, products containing aspartame needs to be avoided. A warning is required on all food products sweetened with aspartame, typically found in small print near the ingredient list:

PHENYLKETONURICS: Contains Phenylalanine

FAQs for Young Children

Children are very observant and will likely notice quickly that your child with PKU is eating different food. Also, while other children may share, it's important to teach your child with PKU that he or she cannot share from his or her friends and classmates. This will likely create questions from these curious children. Prepare your child for these questions by role playing and practicing so when he or she is asked, your child will be ready for it.

Q.Why can't you eat what we are eating?
A. "My body does not break down protein, so I have certain foods I cannot eat. My formula and diet are all a part of that. It keeps me healthy and I feel much better when I stick to it. Protein is kind of [bad for] me, so I have to plan my meals in advance. It takes some time, but in the long run it keeps me happy and healthy." (Patient Perspective)

Q. Are you allergic?
A. Sort of, but if I eat something that I shouldn't, it won't make me sick right away. But later, it could make me very sick.

Q. What happens if you eat something you aren't supposed to?
A. It might not do anything right away, but it might make me grumpy and have a hard time listening and paying attention in class later.

Q. What are you drinking? Why do you always drink that?
A. That's my special drink that gives me the good stuff you would get from food I can't eat. I drink it a lot so I can be strong and healthy.

Q. What kind of treats can you have?
A. I can have lollipops, popsicles and some gum.

Q. Are you sad that you can't eat anything you want?
A. Sometimes I wish I could, and sometimes it's hard not to, but I know it's better for me if I don't.

Resources

Educational Games

RED LIGHT!! GREEN LIGHT!!
Many parents use the Traffic Lights example to teach children about the PKU diet. To create a Traffic Light, draw three circles resembling traffic lights on a large poster board or piece of paper and color them green, yellow, and red. Then cut out pictures of many different types of foods from magazines or websites.

To start the game, explain to your child that there are three kinds of foods: "red", "yellow", and "green". These colors are defined as:

- Green = foods that are low in protein/Phe
- Yellow = foods that are only OK in limited quantities
- Red = foods that are high in protein and not on the PKU diet

Work with your child to organize foods according to each color. Once your child understands these ideas, you can create Traffic Light games to improve or test knowledge. New food pictures can create a fun challenge for your child as he or she figures out where each new food belongs on the traffic light, or you can test your child's recognition by placing a "red" food on the green light, or vice versa, and asking which of the foods does not belong.

The Traffic Light may also help you talk about diet choices with your child. Children familiar with the Traffic Light will readily understand what a "green" food or "red" food is, and this offers a way for parents to say no to foods without using the word "no" constantly. Some parents may also choose to refer to foods as "low Phe" and "high Phe" foods for this same reason.

Low Protein Grocery Shopping Tips

Low protein food offerings have also expanded significantly. Specially formulated breads, pastas, non-meat burgers, cheeses, muffins and cookies are now available, among others, and low protein food companies continue to offer new options for people with PKU.

There are also less expensive low protein foods that are available off the shelves such as those listed below.

PRODUCT	PRICE	PHE
Sunbelt fruit & grain cereal bars (blueberry/strawberry)	$2.00 per box	45-55 mg per bar
Sensible Portions Veggie Straws	$6.00 per 7oz bag	32 mg per serving (38 straws)
Sandwich Mate cheese slices	$1.50 per pack (16 slices)	24 mg per slice
Sun Luck Rice Sticks	~$2.00 per bag	35 mg per serving (1/4 of bag)
KAME bean threads	~$1 per bag	3 mg per 56 gram serving (uncooked)
Pepperidge Farm Very-thin sliced white bread	~$4.20 per loaf	67 mg per slice
Turtle Mountain So Delicious Coconut Milk Yogurt	~1.80 per 6oz	28 mg per 6oz

Resources

The NPKUA website (www.npkua.org) also has a list of low Phe foods available at Trader Joe's and Whole Foods under the PKU Management Tab at Living with PKU.

Support and Informational Resources

National PKU Alliance – www.npkua.org; 1-877-NPKUA-22
The National PKU Alliance (NPKUA) works to improve the lives of individuals and families associated with PKU through research, support, education and advocacy, while ultimately seeking a cure. The NPKUA is the first national non-profit organization to unite adults, families, statewide organizations, the medical community and PKU-friendly businesses under one umbrella organization.

The NPKUA website has links to resources that can help PKU patients and their families living with and managing their PKU, including information about PKU-friendly restaurants, travel destinations and every day information that can support you and your child manage PKU.

State and Regional PKU Organizations
For more information on PKU organizations by state, visit the NPKUA website at www.npkua.org. You can find state organizations listed in the PKU Resources brochure, under the Living with PKU tab.

NORD (National Organization for Rare Diseases) — www.rarediseases.org; 1-800-999-6673
NORD is a not-for-profit health agency dedicated to the identification, treatment, and cure of rare "orphan diseases" such as PKU through education, advocacy, research, and patient services programs.

Patient Power – www.patientpower.info; 1-877-232-5445
Patient Power® is a series of online radio and video programs for patients hosted by Andrew Schorr, a 13-year Leukemia survivor and patient advocate. The programs feature renowned medical experts and inspiring patients discussing an array of chronic medical conditions including PKU. The end goal is patient empowerment.

Social Networking Sites
Social networking sites such as **Facebook** *can offer people with PKU the chance to connect with other PKU patients across the world for tips and support. In addition, local PKU organizations may have Facebook groups that you can join to stay up to date on what is going on in your area. To join, go to www.facebook.com.*

PKU Listserv - Listserv@Listserve.Emory.Edu
The PKU Listserv is a great place for families of a child with PKU, persons with PKU, and professionals involved in PKU treatment to share their ideas and concerns with others. This list provides an easy way for people all over the world to come together and communicate with one another about PKU. If you are interested in joining the listserv, send an email request including your name and email
address to: macpku@verizon.net.

PKU News – www.pkunews.org; (206) 525-8140
National PKU News is a non-profit organization located in Seattle, Washington dedicated to providing up-to-date, accurate news and information to families and professionals dealing with PKU. Since 1989, it has provided a 16-page newsletter three times yearly, and also has available widely used resources for PKU families including a food list, 2 cookbooks, and a children's book.

Resources

PKU.com – www.pku.com
Offered by BioMarin, PKU.com provides comprehensive information about PKU and a place to meet others with PKU. Log on today to join the discussion and to make new PKU friends.

My PKU Toolkit – www.myPKUtoolkit.com
A website created by Applied Nutrition Corp. and Children's Hospital Boston that includes diet tips and model forms for self-management and advocacy for downloading.

PKU Online – www.citt.ufl.edu/team/PKU/beta/index.html
This website was developed by staff members from the Division of Genetics at the University of Florida to teach children about PKU management through interactive games and stories. A section for parents is available as well.

Your Genes Your Health – www.ygyh.org
The Your Genes Your Health webpage focuses on several genetic conditions (including PKU) with explanations and illustrations of the genetics of the condition, incidence, inheritance, symptoms, testing/screening, living with the condition, and treatment.

Wrightslaw – www.wrightslaw.com
This website is devoted to providing information about education law and how to advocate for individuals with disabilities.

Formula and Low-Protein Food Companies and Retailers

Abbott Nutrition – abbottnutrition.com; 1-800-227-5767
Abbott Nutrition provides medical nutritional products for the management of PKU, including Phenex-1 for infants and toddlers and Phenex-2 for children and adults.

Applied Nutrition – www.dietforlife.com; 1-800-605-0410
Applied Nutrition provides medical food for the management of PKU, featuring the Phenyl-Ade line of PKU formulas. Applied Nutrition is also home to Maddy's low protein foods.

Cambrooke Foods – www.cambrookefoods.com; 1-866 4 LOW PRO
Cambrooke Foods provides low protein food and metabolic formula products; products are available to be ordered online and delivered directly to your home.

Dietary Specialties – www.dietspec.com; 1-888-640-2800
Dietary Specialties provides low protein food products for the dietary management of metabolic disorders and medical conditions requiring low protein diets.

Ener-G Foods – www.ener-g.com; 1-800-331-5222
Ener-G Foods provides low protein food products for the dietary management of metabolic disorders and medical conditions requiring low protein diets.

Lil's Dietary Specialty Shop: www.lilsdietary.com; 773-239-0355
Lil's Dietary Specialty Shop is a Chicago-area dietary food store that carries low-protein products. Products can be ordered online and delivered directly to your home.

Resources

Mead Johnson Nutrition: www.mjn.com; 1-800-BABY123
Mead Johnson offers medical formulas for inborn errors of metabolism, such as PKU, including the Phenyl-Free line of medical formulas.

Med-Diet: www.med-diet.com; 1-800-MED-DIET
Med-Diet offers direct to home delivery of medical foods for conditions such as PKU, including their own line of broths and sauces.

Nutricia – www.myspecialdiet.com; 1-800-365-7354
Nutricia created a website devoted to the management of a metabolic diet. It includes an on-line store to purchase a range of medical foods, formulas and low protein foods, as well as cookbooks and software for diet management.
PKU Perspectives – www.pkuperspectives.com; 1-866-PKU-FOOD
PKU Perspectives provides low protein food products for the dietary management of metabolic disorders and medical conditions requiring low protein diets.

Taste Connections – www.tasteconnections.com; 310-371-8861
Taste Connections provides low protein food products for the dietary management of metabolic disorders and medical conditions requiring low protein diets.

Vitaflo – www.vitaflousa.com; 1-888-VITAFLO
Vitaflo provides a range of flexible medical food products for those with PKU, including ready-to-drink medical formula and other convenience optiosn.

Pharmaceutical Company Resources

KUVAN®

BioMarin – www.bmrn.com, www.kuvan.com; 1-866-906-6100
BioMarin commercializes KUVAN®, which is approved to reduce blood PHE levels in patients with hyperphenylalaninemia (HPA) due to tetra-hydrobiopterin- (BH4-) responsive PKU. Call BioMarin Patient and Physician Support (BPPS) to find out about Kuvan's free 30-day trial: 1-877-MY-KUVAN (1-877-695-8826), or email bpps@bmrn.com.

Large Neutral Amino Acids (LNAAS)

The companies below provide Large Neutral Amino Acid (LNAA) formulas for patients with PKU by clinic authorization:

Applied Nutrition: PheBLOC
www.medicalfood.com; 1-800-605-0410

Nutricia: Lanaflex
www.shsna.com; 1-800-365-7354

Solace Nutrition: PreKUnil® and NeoPhe®
www.solacenutrition.com; 1-888-8-SOLACE

Resources

Phe/Protein Food Reference Guides

Emory University's PKU Booklet is a pocket-sized comprehensive food list on the Phe exchange system that includes information about the Phe content in foods, household measurement information, serving sizes, calories, protein and exchanges. There is also room for a favorite food log and to keep notes. Order form is available at http://genetics.emory.edu/docs/Nutrition%20Docs/PKUorderform9-29-05.pdf

Virginia Schuett's "Low Protein Food List"
This food list is available in a 186-page book as well as for computers and handheld devices such as iPods and iPhones. Either format provides Phe, protein and calorie content of more than 3,000 foods. It includes tabbed chapters for Medical Foods, Baby Foods, Fruits & Vegetables, Beverages, Soups, Breakfast Foods, Low Protein Products, Sauces, Fats & Condiments, Grain Products, Crackers & Snacks, Sweets, Baking Ingredients, Convenience Foods, and Very High Protein Foods. Data sources include USDA and manufacturer's data. An index for the book can be downloaded from the National PKU News website, pkunews.org, in the Diet-Related Information section. Additions and corrections to the book are posted in the same section of the website. It is available to order at www.myspecialdiet.com and http://www.pkunews.org/forms/lowproFoodList.htm.

DietWell Application for iPhone and iPads
DietWell™ for PKU app for the iPhone®, iPod touch®, and iPad™ is designed to assist families and individuals in the dietary management of PKU. This app guides you through your meals, snacks, and formula intake by offering phenylalanine (phe), dietary protein, formula protein equivalent (P.E.), and calorie information for over 7500 food items, specially manufactured low protein food products, metabolic formula products, and Cambrooke recipes along with your Kuvan® intake, blood levels and wellness – all at the touch of your fingertips.

Cookbooks and Recipe Resources

WEBSITES FOR PKU RECIPES

Cook For Love: www.cookforlove.org
Cook for Love is a culinary non-for-profit dedicated to the PKU community. Its website and resources provide people with step-by-step instructions and videos for making low protein recipes. Cook for Love's mission is to empower members of the PKU community to improve their health through cooking and education.

Minnesota PKU Foundation: www.mnpku.org/dietrecipe
The Minnesota PKU Foundation is a nonprofit organization that promotes research and the welfare of PKU patients and their families. The website offers recipes for low protein cooking.

PKU of Illinois Foundation: The PKU Organization of Illinois is committed to the support of appropriate research initiatives to better understand PKU and eventually find a cure. Find tasty recipes at on their website at www.pkuil.org/recipes.

Resources

COOKBOOKS FOR PKU RECIPES

Virginia Schuett: Author Virginia Schuett has three PKU-friendly cookbooks available from Nutricia North America via www.myspecialdiet.com. They are

- "Apples to Zucchini: A Collection of Favorite Low Protein Recipes"
  This cookbook contains 562 healthy and delicious recipes in 12 chapters: Salad Celebrations; Soup's On; Bread and Beyond; Vegetables for All Seasons; Rice from East to West; Pasta, Please!; Hot Off the Grill; Where's the Beef?; The Adventurous Cook; Company's Coming; Sweet Delights; And Everything Else. The book's focus is on using naturally low protein fruits and vegetables, with minimal use of special brand-name low protein products.

- "Low Protein Cookery for PKU"
  This "classic" cookbook with over 450 recipes dates from 1977, but is still as relevant today as it was when it was first published. Hundreds of family-favorite recipes grace its pages, utilizing only "basic" special low protein ingredients and easy to find grocery store ingredients. Extensive Helpful Hints and Everyday Tips sections emphasize simple ideas to make the diet more easily managed, especially for younger children (which was the focus of treatment at the time the book was created). The recipes will appeal to a wide age range, from young children to adults and facilitate integration of the diet into normal family eating routines.

Gina Valente:

- "Gina Cooks Low Pro"
  Gina's cookbook gives suggestions and ideas for easy-to-make, quick and healthy low protein recipes. Example recipes include ice cream, pizza, burgers, mac and cheese, meatless balls, tortillas, nacho chips, pasta salad, pizzelles, chocolate cake, puddings and much more. Gina creates her recipes right from the grocery store using a few low pro foods (such as low pro pasta and low pro mixes). She suggests how to incorpo rate fiber, flax, omegas and other nutrients to the PKU diet that can otherwise be deficient. Futhermore, all of Gina's recipes have been reviewed and calculated by a registered dietitian for protein, PHE and calorie counts. A special offer of $16.99 plus S&H has been extended to those who use the code: NPKUA.ginacookslopro. T o order, email: ginacooks@optonline.net

Resources

MACPAD: The Mid-Atlantic Connection for PKU and Allied Disorders (MACPAD) is a non-profit organization dedicated to improving the health and well being of individuals and families affected by PKU and related metabolic disorders. MACPAD offers two cookbooks special for PKU diets:

- "Creative Family Cooking"
  Blood, sweat and possibly tears have gone into each creation in this collection of delicious recipes from people from the PKU community. Some people are blessed with the ability to create new recipes, others can adapt existing recipes and others have the skill to know what recipes will appeal to those with PKU. Some of the recipes are basic and others more involved, but each brings an individual accomplishment and adds to the knowledge of dealing with the sometimes challenging dietary regime. The name of each chef is featured along with the recipe. Available at www.MACPAD.org for $10.

- "Family Friendly PKU Recipes"
  The driving force behind this, their second cookbook, was to find and adapt recipes that can be shared with the entire family, including family friendly recipes to compliment the low protein components. Each recipe was tasted and tested by MACPAD members and friends. Inside you will find sections like Best Baked Goods; Soups, Salads and Salsas; Sensational Salads; Enticing Entrees; Vivacious Vegetables; Fantastic Fruits and Sweets and Snacks. This cookbook is also available at www.MACPAD.org for $18.

Ordering Information

Scales
Most practitioners and PKU patients recommend gram scales made by Ohaus or Tanita. To purchase scales, many PKU patients recommend the website www.oldwillknottscales.com or eBay, but if purchasing from eBay, make sure the scale is new, not used.

Any scale you purchase should have the following features:
- The ability to weigh in grams in 1 gram units
- The ability to weigh up to 1,000 grams
- A tare function that allows you to zero out the weight of any container placed on the scale (before you add any food or formula) to get an accurate measurement of only the food or formula

Lancet Device
You should maintain an adequate number of lancet devices for blood sampling. Using a 21-gauge, trigger loaded lancet pen with settings for different depth options for the stick is recommended for PKU patients. These can be ordered through your local drug store.