

# An Educator's Guide to PKU

*For Educators of Students who have Phenylketonuria (PKU)*



**Boston Children's Hospital**  
Until every child is well™



“There is a big learning curve for teachers when they have a student with PKU in their classroom for the first time. It would be helpful to have information about PKU to pass onto teachers so the learning curve doesn’t have to be so big each year.”

– Vice Principal of an elementary/middle school with three students with PKU



## Dear Parents and Educators,

This guide to PKU has been created for teachers to help support children with PKU at school. Both the parent and educator communities have expressed a need for a resource like this and we’ve heard you loud and clear. We’ve done some research on our own, but have also talked to parents and teachers like you to find out what you need and want to know about PKU. The guide we’ve developed includes information about the low-Phe diet, new research on the most common learning challenges in PKU and how they become apparent in the classroom, guidance for educators on what they can do to help, and space for you to add notes about the child with PKU in your life.

Here are some helpful tips for using this guide:

- This guide should be reviewed in a meeting between parents, teachers, and any other school personnel involved in the education and care of a child with PKU. This can help to align parents and educators on the educational and developmental goals for the child, as well as open the lines of communication for the year.
- If possible, have this meeting before the school year starts or even at the end of the previous school year. Teachers believe that this would help them to devote more time and attention to the needs of a child with PKU before the other students arrive.

We hope you will find this educator’s guide both informative and useful and see it as a helpful resource in supporting children with PKU at school!

Best Wishes,

**Joanna Logue, MA**, Emerson College

**Susan Waisbren, PhD**, Boston Children’s Hospital

## PKU at a Glance

Phenylketonuria, or PKU, is an inherited metabolic disorder that affects about 1 in every 10,000 people in the United States.

Individuals with PKU are unable to break down phenylalanine, or Phe, which is an amino acid found in all protein. For this reason, people with PKU must limit the amount of protein they consume. In order to have a healthy diet, they must drink a special formula that contains all the parts of protein, except for Phe. This diet can be difficult to maintain, which presents many challenges for those with PKU and their families.

When individuals with PKU do consume Phe, it builds up in their blood. High levels of Phe in the blood affect cognitive functioning and create difficulties in learning, behavior, and social interactions.

The good news is that PKU is diagnosed through newborn screening and a simple blood test helps to keep track of blood Phe levels over time. This, along with the availability of a low-Phe diet, makes it possible for individuals with PKU to live normal and healthy lives.



**A special formula helps children with PKU get needed nutrients and calories**

*– Drawn by a 5th grader with PKU*

## The PKU Diet

### Foods to be Avoided

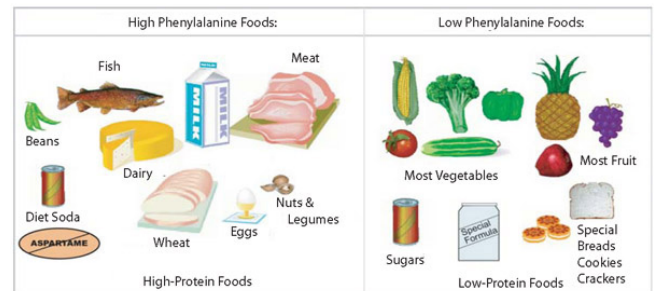
Common foods and drinks that contain high levels of Phe and should be avoided include:

- Meat
- Turkey
- Milk
- Cheese
- Eggs
- Nuts
- Soy
- Chicken
- Fish
- Ice cream
- Yogurt
- Beans
- Peanut butter
- NutraSweet

### Allowable Foods

Common foods and drinks that contain lower levels of Phe and are allowed in measured amounts include:

- Fruits
- Vegetables
- Potato chips
- Fruit juices
- Cereal
- Some candy



Children with PKU can always have an apple. It's a Phe-free food!

Special foods containing little or no Phe are produced specifically for individuals with PKU. These include low-protein breads, baked goods, pastas, and even low-protein cheese and meat alternatives.

**There is wide variability among children with PKU in their dietary preferences and nutritional needs. It is important to communicate with a child's parents to make sure he or she is eating the right foods and enough food while at school.**

## Common Neuropsychological Deficits in PKU

### 1) Executive function

- Attention
- Working memory
- Planning
- Organization

Thought to be related to modest reductions in the prefrontal cortex

### 2) Information processing

Thought to be related to reduced myelin in the brain

### 3) Visual-motor coordination

Thought to be related to abnormal neurotransmitter levels in the brain

Sometimes associated with PKU are:

- Increased stress and depression
- Social anxiety, withdrawal, and negative self image

Prevalence of symptoms and learning deficits in a sample of 55 children with early-treated PKU at one metabolic center:

ADHD	Behavior Problems	Memory Deficits
53%	52%	50%

Attention Deficits	Visual-motor Coordination Deficits
37%	42%

**Although the prevalence of these symptoms and learning deficits is high, the average IQ in this study sample is 99 ±14. The average IQ for the general population is 100 ±15.**

**“Staying focused and memorizing facts were difficult for her. We found that this was related to her Phe levels.”**

**– Academic tutor of an 8th grader with PKU**

## PKU in the Classroom

The learning deficits in PKU become apparent in the classroom environment in different ways. These might include:

- ADHD-like behavior, such as hyperactivity and inattention
- Difficulty with math and reading comprehension
- Trouble multi-tasking and following multi-step instructions
- Difficulty finishing tests or assignments in the time allotted and in keeping up with note-taking
- Difficulty understanding charts and diagrams
- Trouble comprehending orally presented materials
- Low energy and fatigue
- Struggling to pack a backpack or fasten a jacket
- Sitting alone at lunch or recess
- Worrying to the point of becoming sick

**Research shows that the symptoms of PKU become more pronounced when blood Phe levels are too high. For this reason, it is important to notify parents when these classroom behaviors occur so they can take steps to lower their child’s Phe levels.**

“The 504 plan changed her life.”

– Mother of a 5th grader with PKU

“Her IEP is like a safety net. It gives her support when things are tough.”

– Mother of a 10th grader with PKU



## Section 504 and IEP

Because children with PKU experience many challenges at school, there are plans available to make sure they have the best chance of succeeding in the classroom.

### Section 504 Plan

A Section 504 Plan ensures that the needs of children with PKU are met as adequately as those of other children. The list of school accommodations to achieve this goal might include:

- Extended time on tests or assignments
- Oral tests and visual aids
- Rearranging class schedules
- Providing low-Phe foods in the school cafeteria
- Allowing a child with PKU to keep low-Phe formula at his or her desk

To learn more about Section 504 plans, visit:  
[http://www.nldline.com/iep\\_vs\\_504.htm](http://www.nldline.com/iep_vs_504.htm)

### Individual Education Plan (IEP)

An IEP is a written statement of an educational program designed to meet a child's individual needs. An IEP sets reasonable and attainable learning goals for a child with PKU.

To learn more about IEPs, visit:  
[http://www.nldline.com/iep\\_vs\\_504.htm](http://www.nldline.com/iep_vs_504.htm)

**Children with PKU have a legal right to these plans and it is important that they be followed at school. Teachers should work with parents, the metabolic health care team, school psychologist, school nurse, and other school personnel to decide on a plan that meets the specific needs of each child with PKU.**

## What Educators Can Do to Help

Beyond the IEP and 504 Plan, there are other ways that educators can help children with PKU.

### Academics and Learning

- **Set high expectations for a child with PKU.** Research shows that this is parents' biggest request of teachers. Not all children with PKU experience learning challenges.
- **At the same time, be patient and flexible.** A child's academic performance can change depending on fluctuating Phe levels. If you notice any changes, tell parents so they can take steps to adjust the child's intake of Phe.
- Some research suggests that slow information processing is the underlying cause of what appears to be attention deficits in PKU. For this reason, speaking slowly and combining verbal explanations with visual or written information can be helpful for children with PKU.

### Food, Parties, and Field Trips

- Low-Phe foods and formula will often be sent to school as part of a child's lunch or snack. Encourage the child to eat these foods and allow him or her to drink formula throughout the school day.
- Communicate with parents and school administration to know if the school has purchased low-Phe foods to be made available in the cafeteria.
- Information on low protein foods that schools can purchase is available through Cambrooke Foods' School Lunch Program. To learn more, visit: <http://www.cambrookefoods.com/information/index.php#lunchline>

**“At first, he was embarrassed and kept his PKU a secret from the other children. This makes it harder for them to understand and accept it now that they know.”**

**– Mother of a 7th grader with PKU**



- Ask parents specifically what foods a child can and cannot have.
- Ask parents for a “PKU food list” to help you understand the child’s diet.
- Tell parents when there is a class party so they can provide a low-Phe treat.
- Ask parents if they want to send in a batch of low-Phe treats that can be frozen so they are available for class celebrations.
- Serve fresh fruit and vegetables instead of baked goods at parties. This can make food choices easier for children with PKU.
- **If a child with PKU eats food with Phe, it is not a medical emergency.** Just be sure to notify the parent at the end of the day.
- **If a child doesn’t finish his or her food or formula during the school day or eats food with Phe, tell the parent.** This is important so they can adjust the child’s dinner for that night or meals for the next day to balance out the child’s Phe intake and maintain the child’s Phe levels within the recommended range.

- Some children take a new prescription drug, known as Kuvan (sapropterin dihydrochloride), to treat their PKU. Changes in behavior could indicate the drug is not working, so it is important to notify parents.
- Psychologists in many hospital metabolic clinics are available to discuss behavioral changes and symptoms in children with PKU.
- Organize class activities that highlight the differences among people to show that there are many things, not just PKU, that make people different.
- Allow older children with PKU to write a report or give a presentation to classmates about the disorder. This can help the child to feel confident among peers.
- **Treat a child with PKU as a normal, healthy member of the class.**

Communicating with parents is important in helping children with PKU to succeed in the classroom. As an educator, you are a parent’s eyes and ears during the school day and play an important role in the health and well being of the child.

### **Behavior and Social Interactions**

- Be on the lookout for the behavioral symptoms of PKU.
- Behavioral patterns can change depending on fluctuating Phe levels. If you notice any changes, tell parents so they can take steps to adjust the child’s Phe intake.

**“The school has been very supportive. Teachers let her retake tests and make a point of using visual aids and analogies to help her understand the material.”**

**– Mother of a 10th grader with PKU**

## Additional Resources

### The New England Consortium of Metabolic Programs

This website features information and resources from a variety of health professionals aimed at providing care for people with metabolic disorders.

<http://newenglandconsortium.org/>

### The New England Connection for PKU and Allied Disorders

This website includes information, resources and activities to support and benefit individuals with PKU and their families in New England.

<http://www.necpad.org/>

### National PKU Alliance

NPKUA is a collaboration of PKU community members who support efforts to raise PKU awareness, promote advocacy and education, and search for a cure for PKU.

[www.npkua.org/](http://www.npkua.org/)

### National PKU News

This website provides up-to-date and accurate news and information for families and professionals who deal with PKU.

[www.pkunews.org/](http://www.pkunews.org/)

### GEMSS – Genetics Education Materials for School Success

GEMSS features information about genetic disorders, including PKU. It is designed for parents and teachers to provide them with the information and resources they need to help children with genetic conditions to succeed at school.

<http://www.gemssforschools.org/>





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- Parents and educators of children with PKU
- Elementary and middle school teachers
- Freedom Baird, MS, Web Consultant, Boston Children's Hospital



### Parent/Guardian Notes

In this section, parents or guardians can make notes or recommendations about their child with PKU that might be helpful to share with teachers and other school personnel.

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**Teacher Notes**

In this section, teachers can make notes or recommendations about a child with PKU that might be helpful to share with other teachers and school personnel.

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## **An Educator's Guide to PKU**

Created by:

Joanna C. Logue, MA

Emerson College, Health Communication Program

Supervisor: Timothy Edgar, PhD

Preceptor: Susan Waisbren, PhD

This work was created in collaboration with the Metabolism Program at Boston Children's Hospital

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For additional information, please contact:

Susan Waisbren, PhD

Metabolism Program, Division of Genetics

Boston Children's Hospital

1 Autumn Street

Boston, MA 02215

617-355-7346

[childrenshospital.org](http://childrenshospital.org)



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