# The ABC's of PKU

(Phenylketonuria)

# What Early Childhood Educators and School Administrators Need to Know

***Because Knowledge Leads to Better Health***

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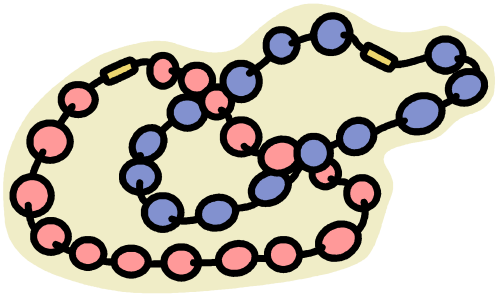
# What is PKU?

PKU is a genetic disorder of metabolism affecting approximately 1 of every 15,000 babies born in Canada. PKU patients lack the enzyme in the liver needed to turn phenylalanine (phe), an amino acid of protein, into tyrosine. This results in an accumulation of phe in the blood which crosses into the brain and is **toxic**, causing various degrees of mental deficiency and neurological issues.



**Protein is made up of a chain of amino acids similar to a beaded necklace. Phenylalanine is one of those beads (or amino acids).**

In someone <u>without</u> PKU, a small portion of the phe eaten each day is used for growth and the leftover phe is turned into Tyrosine. Tyrosine is used to make the neurotransmitters norepinephrine and epinephrine, which relay nervous system messages throughout the body. Since only a small portion of phe is used for body growth, and the remaining phe can't be broken down into Tyrosine, **the diet of a PKU patient needs to be calculated very precisely.** PKU patients can only eat the amount of phe needed for growth each day, but no more so that a buildup of Phe in the body doesn't occur. Blood-Phe levels are monitored regularly (usually once every two weeks for school-age children) and the diet is adjusted by a specially trained dietitian according to the result of the blood test.

PKU patients in Canada attend regular visits to a specialized clinic and the management of their condition is monitored by a team that can include doctors, dietitians, nurses, psychologists, genetic counselors and social workers (different clinics have different resources available.) **Parents/guardians of PKU children and PKU patients themselves quickly become experts in this rare disorder, so please trust that what they are asking you to do is essential for the health and well-being of the PKU student in your care!**

# How is PKU Managed?

***The Formula***

PKU patients drink a special formula every day, and will do so for the rest of their lives. This formula provides the now essential amino acid Tyrosine, since the PKU patient cannot produce tyrosine, as well as other vitamins, minerals and nutrients otherwise lacking from the severe phe-restricted diet. **This formula is essential for proper growth, blood-phe levels and brain function.** It is crucial that 100% of the prescribed formula is taken every day, ideally spread evenly throughout the day. This formula often requires refrigeration and PKU patients sometimes try to hide or throw away their formula in fear of what their peers will think. It is essential that you make accommodations for your student to drink this special formula as instructed by the parent/guardian, and provide a safe environment.

***Protein Intake***

PKU patients must follow a severe, phe-restricted diet. Phe is counted in milligrams (mg) or in exchanges (1 exchange = 15mg of phe). Some patients are able to count grams of protein but still take in a very restricted amount of protein.

The food labeling rules of Health Canada require the listing of the amount of protein in grams but do not require the listing of the amount of phe. Parents/guardians must research to find out exactly how much Phe is in any particular food.

Every patient has a different phe tolerance – that is, the amount they can ingest each day without causing dangerous blood-phe levels. Some diets are as low as 10 exchanges (150mg) of phe each day, or even lower. Here are some examples of the phe in various foods.

## Lower protein Foods

1 medium (180 gram) apple =

**0.6 exchanges or 9 mg of phe**

½ Cup (40 grams) Broccoli =

**2.2 exchanges or 33mg of phe**

½ Cup (50 grams) Cauliflower =

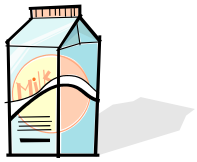
**2.3 exchanges or 34.5mg of phe**

1 medium (114 gram) Banana

**2.9 exchanges or 43.5mg of phe**

## Higher Protein Foods (off limits)

1 fluid ounce Milk =



**3.3 exchanges or 50mg of phe**

¼ Cup (45 grams) Chocolate Chips =

**9.3 exchanges or 140mg of phe**

1 medium (50 grams) Egg

**20.0 exchanges or 300mg of phe**

1 tbsp. (9 grams) Peanuts

**7.3 exchanges or 110mg of phe**

As you can see, patients must comply with a very restricted diet which usually consists only of fruit, vegetables and specially produced low protein foods which are not available in regular grocery stores. **PKU patients cannot eat any MEAT, FISH, EGGS, SOY, DAIRY, NUTS, LENTILS OR SEEDS and very limited amounts of GRAIN and FLOUR products, if any.**

***Calculating Phe***

The parent/guardian will weigh all the food to calculate the exact phe amounts before sending the packed lunch to school. **However, it is what is *ingested* that matters; therefore it is essential that**

**leftovers are sent home so the phe intake can be recalculated.** If you and the parents are comfortable doing so, the parent can provide you with a scale so that you can be more involved with the management. This may allow for some flexibility (for example, if another student brings fruits or vegetables to share with the class, you can weigh an appropriate amount for the PKU patient.) The scale is also a great tool that you can incorporate into the classroom learning!

***Recording Food Intake***

**Parents/guardians of PKU children need to record daily everything that is eaten by the patient.** This is essential for both the day to day management of the disorder, and as a reference to determine what has caused a high blood-phe level.

Here is a sample of a daily food intake record:

| | Food | Amount | Exch. | Mg phe |
|---|---|---|---|---|
| Breakfast | Pancake w/ 30ml Rice Dream | 1 | 0.2 | 3 |
| | Apple Juice | 4 fl.oz | 0.0 | 0 |
| | *Formula* | | | |
| | | | | |
| Snack | Gerber Juicy Treats | 1 pack | 0.33 | 5 |
| | | | | |
| Lunch | Fries | 24 gram | 2.0 | 30 |
| | Ketchup | ½ tbsp | 0.25 | 3.75 |
| | Baby Dill Pickles | 28 gram | 0.2 | 3 |
| | Apple Juice | 4 fl.oz | 0.0 | 0 |
| | *Formula w/nap* | | | |
| | | | | |
| Snack | SunRype FunBites | 1 pack | 0.67 | 10 |
| | Mandarin Orange | 60 gram | 0.86 | 13 |
| | | | | |
| Dinner | Cauliflower | 28 gram | 1.17 | 17.5 |
| | Imitation Cheese Sauce | 2 tbsp | 1.06 | 16 |
| | Rice Dream | 45 ml | 0.26 | 4 |
| | *Formula* | **Total:** | **7.00** | **105.5** |

**It is important to know that the PKU diet tends to be very high in sugar**. Sugar, oils and most fats are phe-free and help provide essential calories (too few calories results in *catabolism*, a breaking down of our muscles that results in high blood-phe level).

**It is also important to know that ASPARTAME is strictly forbidden**, as it is 50% Phe. Anything containing Aspartame (also known as brand names Equal and NutraSweet) is off limits!

Please note that although the PKU patient eats a very different diet, it is prescribed on an individual basis and meets their nutritional needs perfectly as long as all the formula and the right amount of phe is ingested each day!

# What This Means for You

There are four general rules that all caregivers, educators and administrators need to follow in order to protect their PKU student:

1. **NEVER give the student anything to eat other than what is sent in their lunch, unless you have pre-approval from the parent;**
2. **NEVER allow PKU patients to share their lunch with others, or eat food from another student's lunch;**
3. **ALWAYS ensure that the food left over from the student's lunch is sent home so that the parents/guardians can record exactly what was eaten and make adjustments as necessary;**
4. **ALWAYS tell the parents/guardians as soon as possible (immediately or at the end of the school day) if something other than the packed lunch was eaten.**

Those rules are essential for the health and well-being of your student. However, there are other things you are able to do to help the patient feel positive about their diet:

1. **INFORM** all the parents/guardians of the classroom that there is a student with special dietary requirements and request that they always give at least 24 hours of notice before bringing special treats for the class to share (birthday cupcakes, Halloween treats, etc.);
2. **CONSIDER** distributing the list of safe treats provided by the parent/guardian to the other parents so that they can send treats the PKU student can share;
3. **STORE** some low-phe or phe-free treats (pre-approved by the parent) to give to the PKU patient should unforeseen treats come into the classroom. **Parents/guardians MUST be advised that a treat has been given before the end of the work day!**
4. **INCLUDE PKU** in your lesson plans whenever possible: point out differences when teaching about the Canada Food Guide; use the scale and examples of the phe calculations in math lessons, teach students that each of them is unique, etc.

These additional efforts are not essential but they will definitely help the student in the long term. One of the biggest risks of the strict PKU diet is the lack of compliance as patients age, and any time a patient feels embarrassed, left out, ostracized or ridiculed because of their PKU will increase the likelihood of non-compliance.

# PKU Effects on School Performance

Your PKU student should be treated like any other student, with the exceptions of the dietary requirements. Due to the miracle of Newborn Screening Programs, most patients in Canada are diagnosed within a couple weeks of birth and are able to avoid the serious mental defects associated with uncontrolled PKU. However, your student may be late-diagnosed, meaning that some brain damage occurred before the proper treatment was implemented, or may have neurocognitive or behavioural issues related to high Phe levels. **High Phe levels are unavoidable in some instances (growth spurts and illness, for instance) and do not reflect a poor job by the PKU parent/guardian!**

PKU patients are at higher risk than their peers for certain traits that can affect their performance at school. **As their teacher, you may be the first to notice one or more of the following symptoms and it is important that you communicate any concerns to the parents/guardians as soon as possible.** PKU patients are at higher risk of:

- Depression
- Anxiety
- Panic Attacks
- Agoraphobia
- Poor working memory
- Poor ability to focus
- Difficulty planning and organizing
- ADD or similar symptoms
- Reduced IQ
- Slow Processing Speed
- Poor impulse control

If a patient develops one or more of these conditions, they may require extra help in school, an Individual Education Plan, or other form of aid. Whether resources are available varies by province and district, but CanPKU can support your request for extra assistance by providing information and research. If you need help in this regard, please email us at: info@canpku.org.

# Frequently Asked Questions

***What should I do if the PKU student eats something he/she shouldn't (i.e. food from another student or something given accidentally)?*** It is crucial that you let the parents/guardians know as soon as possible as their whole diet for the rest of the day, including their formula, may have to be altered. If you are able to call the parent/guardian immediately, please do so; otherwise please contact them after school hours. Direct communication is key (i.e. it is not sufficient to simply send a note home with the student.)

***What should I do if the PKU student spills or does not want to drink his/her formula?*** Again, it is very important for the parents/guardians to find out as soon as possible as they will have to make adjustments. Please try to estimate how much of the formula was lost (in the event of spillage) and ensure the student drinks the rest.

If the student is unwilling to drink their formula or eat their pre-packed lunch, please encourage them to do so. Most PKU patients understand the importance of their diet and don't start to rebel until they get closer to their teen years, but even a small amount of teasing from their peers can have a very negative effect. If the child refuses despite your encouragement please communicate directly with the parents/guardians about this serious issue as soon as possible. Even a week of mismanagement can result in noticeable behavioural or neurocognitive effects such as those listed above.

***What should I do if the PKU student is feeling left out of a class treat and I have nothing suitable to give them instead? Can I give them just a little bit?*** NO. There are two major consequences of giving 'no' foods to a PKU patient. The primary concern is that too much phe can cause damage to the brain. The secondary concern is that you may be introducing a 'no' food for the first time. It is extremely difficult for anyone to resist foods that they like so PKU patients are much more likely to stay compliant with their diet if they never taste the foods that they are not allowed to have. The idea that 'a little bit won't hurt' is untrue. A little bit, in fact, may be setting the patient up for non-compliance and serious consequences later on in life.

***Where can I get more information?*** Canadian PKU and Allied Disorders Inc. is a non-profit organization dedicated to helping families and professionals who deal with PKU. Please visit our website at www.canpku.org and feel free to contact anyone listed on the 'Contact Us' page.

***What should I tell parents are acceptable snacks to bring to class, so that the PKU patient can have some as well?*** This varies by individual patient. We expect the parent/guardian to provide you with a list of foods that are acceptable, as well as the maximum grams allowed, should you be willing to use the scale and weigh out portions. Remember, even if a food is listed below and approved by the parent/guardian you must let the parent/guardian know at the end of the school day that extra food has been consumed, what the food was and how much the PKU student ate!

# *PKU Student Information*

**NAME:** **NICKNAME:**

**BIRTHDATE:** **DAILY PHE INTAKE:**

**DIETARY REQUIREMENTS DURING SCHOOL HOURS:**

Formula quantity and time(s) to be given:

Notes regarding lunch and snacks:

Acceptable replacement treats to have on hand:

**CONTACT INFORMATION:**

Name of Primary Contact:
Phone Number(s):
Email Address:

Name of Secondary Contact:
Phone Number(s):
Email Address:

***Thank you for learning about PKU… Because Knowledge Leads to Better Health!***