Including students with PKU

Teaching 4830 – Strategies for effective inclusion

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27 September 2017
Plan

What is PKU?
- Details of PKU
- Living with PKU

Teachers and PKU
- We’d like you to know...
- PKU and inclusive classrooms
Overview

- PKU (phenylketonuria) is a disorder affecting metabolism.
- About one of every 15,000 children is born with PKU. It is genetically inherited (autosomal recessive) and permanent.
- PKU causes intellectual disability, but this can be avoided by following an extremely restricted diet.
In the U.S., newborns are screened for PKU and other disorders when they are 2 days old: this is the “heel-prick test.” Without this test, there is no way to recognize PKU until brain damage has already occurred.
Proteins & amino acids

Proteins are chains of molecules called amino acids. There are 20 different kinds of amino acids, including phenylalanine, or “phe,” and tyrosine.
In the liver, phenylalanine is converted to tyrosine.

The body uses tyrosine as a building block in lots of things, including dopamine and melanine.
People with PKU lack the PAH enzyme, so phe can’t turn into tyrosine. Instead, it builds up in the blood.

The extra phe clogs brain receptors and interferes with brain development.
The PKU diet

People with PKU must eliminate almost all phe from their diets to keep their blood levels low.

The problem is that any natural protein contains phe. People with PKU still need the other amino acids – and extra tyrosine.
NutraSweet® (aspartame, or Aminosweet®) is 50% phe, so food with NutraSweet must have a warning label.
SuperMilk!

Formulas like Phenyl-Free 2 provide necessary nutrients, but do not contain phe.

The formula is mixed with water, like infant formula. It tastes good but smells terrible, so we use a covered bottle or a child’s “sippy cup.”
Other food

We eat other food to satisfy hunger, and to get the small amount of phe we need. Daniel needs about 300 mg of phe per day: about the amount in one egg.

We use a lot of special recipes for low-phe treats. We get some low-phe food, like no-grain pasta, from nutritionists at UW-Madison.

When we’re away from home, we always pack our own food – even for long trips!
## Recipe comparison

<table>
<thead>
<tr>
<th>“Fast white bread,” <em>(Joy of Cooking)</em></th>
<th>“Sandwich bread”, CookForLove.org</th>
</tr>
</thead>
<tbody>
<tr>
<td>~145 mg phe / slice</td>
<td>22 mg phe / slice</td>
</tr>
<tr>
<td>3 C. flour</td>
<td>(\frac{1}{4}) C. flour</td>
</tr>
<tr>
<td>1 C. water</td>
<td>(\frac{1}{4}) C. water</td>
</tr>
<tr>
<td>2 T. butter</td>
<td>2 T. butter</td>
</tr>
<tr>
<td>1 T. sugar</td>
<td>3 T. honey</td>
</tr>
<tr>
<td>2(\frac{1}{4}) t. yeast</td>
<td>2 t. yeast</td>
</tr>
<tr>
<td>1(\frac{1}{4}) t. salt</td>
<td>2 t. salt</td>
</tr>
<tr>
<td>1(\frac{3}{4}) C. wheat starch</td>
<td>1(\frac{3}{4}) C. wheat starch</td>
</tr>
<tr>
<td>(\frac{2}{3}) C. tapioca starch</td>
<td>2 T. Metamucil</td>
</tr>
<tr>
<td>2 T. Metamucil</td>
<td>2 t. xanthan gum</td>
</tr>
<tr>
<td>2 T. potato flakes</td>
<td>2 T. potato flakes</td>
</tr>
<tr>
<td>1 C. rice milk</td>
<td>1 C. rice milk</td>
</tr>
</tbody>
</table>
Daniel’s daily diet

- **Breakfast**
  - 1 C. Kix, dry (80)

- **Lunch**
  - 2 slices buttered toast (special recipe) (44)
  - 1 apple (5)
  - 1 mini-box Nerds (0)

- **Snack**
  - 4 French twists (44)

- **Dinner**
  - pizza (45)
    - pizza crust (special recipe)
    - marinara sauce
    - pineapples
    - bell peppers
    - shredded imitation cheese
Blood tests

We monitor the amount of phe in Daniel’s blood with a finger prick test, about once every three weeks.

We mail the sample to the Biochemical Genetics Clinic at the Waisman Center at UW-Madison, and they email the results to us.
Common misconceptions

PKU is not the same as:

▶ veganism/vegetarianism
▶ celiac disease
▶ lactose intolerance
▶ a food allergy
▶ food preferences
“[P]atients with PKU may have:
► learning difficulties,
► school problems,
► decreased motivation,
► less social competence,
► irritability,
► hyperactivity,
► mood disorders, and
► poor self esteem.”

Students with PKU, and their parents, are experts on adapting activities for their specific needs. Communicate with them when classroom activities involve food!

You cannot provide low-phe pizza for the class pizza party, but with advance notice, we can. Plan ahead!
Example: teaching the food pyramid
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Image sources

- peptide: http://users.rcn.com/jkimball.ma.ultranet/BiologyPages/P/PrimaryStructure.html
- doublemint: http://pkugirl.blogspot.com/2012/01/aminsweet-is-aspartame.html
- USDA food pyramid: http://www.nal.usda.gov/fnic/Fpyr/pyramid.gif
- PKU food pyramid: http://www.lowprotein.com/food_guide_pyramid.htm