



Eating for Two Nutrition in Maternal PKU

Everyone knows that nutrition is an important part of any pregnancy. But for the woman with PKU, eating correctly takes on even a higher level of importance. In maternal PKU, following the recommended diet is the only way to lower maternal blood phenylalanine levels to the range that is safe for fetal development. Otherwise, the baby has a high chance of having problems, including heart defects, small head size and mental retardation. Thus, women with PKU face enormous pressure to follow the diet correctly.

There are three essential components to the maternal PKU diet: taking the special phenylalanine free formula (often called medical food), restricting phenylalanine intake, and getting enough calories to gain weight normally.

Formula

Formula can be considered "custom made" or modified protein; it provides all the amino acids that are needed for the woman and her developing baby except for the amino acid, phenylalanine, which can not be properly metabolized in PKU. Without the formula, the woman and baby are starved for protein. The formula also provides calories, vitamins and minerals, but these are of secondary importance because there are other means of getting such nutrients. However, in PKU there is no way other than the formula to provide sufficient protein.

The formula has a characteristic taste and odor that comes from protein being supplied as individual amino acids rather than whole protein. For many women, especially those who are returning to diet for pregnancy, drinking the formula is a major challenge. As a resource mother, it is critical to know if the woman is taking all her formula, because without it a woman can not have a successful outcome to her pregnancy, and all other food or support you provide will not matter.

Formula is best used by the body if the woman drinks it throughout the day; usually three or four servings is recommended. The type and amount of formula a woman with PKU needs is determined by her PKU clinic. Formula is prescribed by the doctor or nutritionist to meet protein needs, and because the amount of protein each formula contains differs, so does the total amount prescribed. Under no circumstances should the formula be changed without consulting the medical team. However, if you recognize that the formula is not being consumed, try to identify ways to help the woman accept the formula, and discuss the problem with the PKU clinic staff.

Because of the characteristic taste and odor of the formula, it may be especially difficult to tolerate during the early weeks of pregnancy when nausea and vomiting are common. Women are sometimes reluctant to admit having difficulty with formula intake because they are emotionally overwrought knowing that they can not tolerate the formula and fearing the consequences to the infant. Others may not keep close track of the amount of formula they have consumed, and report taking the amount they have been prescribed. Still others may not understand the importance of taking the formula to achieving metabolic control and proper nutrition.

If taking formula is deemed to be the problem, several options exist:

1. Flavor the formula with juices, syrups, extracts, sugar, or Koolade, to name a few.
2. Drink formula from a covered cup or sports-type bottle to reduce the odor.
3. Chew gum, have hard candy or brush teeth after taking the formula to reduce the aftertaste.
4. Try a different formula (including bars and capsules instead of beverage) this must be discussed with and prescribed by the PKU clinic.

Phenylalanine Restriction

The second component to the diet is restricting phenylalanine. Phenylalanine is one of the amino acids found in all protein, therefore any food that contains protein also contains phenylalanine. The amount of phenylalanine that each woman with PKU can handle (phenylalanine tolerance) varies widely depending on her degree of PKU, her health, and how rapidly the fetus is growing. During periods of rapid fetal growth, phenylalanine tolerance increases because the fetus has a higher demand for phenylalanine in order to build its body tissues. Frequent monitoring of blood phenylalanine levels shows if a woman is getting the right amount of phenylalanine. If her level is <2 mg/dl she may be able to tolerate more, if her level is >6 mg/dl she is most likely getting too much phenylalanine.

There are several methods for counting dietary phenylalanine; some women count exchanges or equivalents, some count milligrams of phenylalanine, some just count "higher" foods such as potato and regular starch, and some count grams of protein. Before meeting with your Resource Daughter, you should talk to the nutritionist at the PKU clinic and learn which method is being used. Regardless of the counting method, the woman with PKU should understand the system, weigh or measure her portions sizes, and keep within the phenylalanine limit she has been prescribed. Keeping a journal of her food and formula intake is the best way for you and the PKU clinic nutritionist to know whether or not the woman understands and is adhering to the diet.

Sufficient Caloric Intake

Women with PKU should gain weight normally during pregnancy despite the fact that they follow restricted diets. The nutritionist or doctor will tell you whether the woman is gaining weight optimally. Weight gain during pregnancy is a predictor of birth outcome in maternal PKU. In the MPKU Collaborative Study, protein intake and weight gain were found to be the most important nutritional variables for determining the health of the baby.

Inadequate weight gain is due to either insufficient protein intake or insufficient caloric intake. Protein and energy intakes are interrelated; adding more calories if the protein intake is low is futile. Insufficient protein intake occurs when the amount of medical food (formula) prescribed is insufficient and/or the woman is having difficulty consuming the proper amount of medical food. Do not easily disregard the possibility that the woman is having difficulty consuming the full amount of medical food if her weight gain is inadequate or her phenylalanine levels are high even if she reports adherence to taking the medical food.

Weight loss will result in high blood phenylalanine levels and needs to be corrected rapidly. Unfortunately, in pregnancy weight loss is often due to nausea and vomiting, making it difficult for the woman to get sufficient calories. Formula is often not tolerated which compromises protein, energy and overall nutrient intake. If usual corrective measures for nausea and vomiting are not working, hospitalization for nutritional repletion may be necessary.

Promoting Weight Gain

Weight gain is important for maintaining blood phenylalanine levels in the desired range and for supporting the growth of the fetus. If the doctor or nutritionist says that the Resource Daughter is not gaining enough weight, do everything you can think of to try to promote weight gain. Given the limitations of the diet, extra calories can be added only from fats, sugar, and low protein specialty products.

Fat.

Make certain the woman is getting adequate fat. Women in the Maternal PKU Collaborative Study tended to have lower than recommended fat intakes. In this age of fat phobia, many women use low fat/non fat salad dressings and other products - these should be avoided. Give a specific number of servings of high fat foods to be eaten each day, rather than general guidelines as her idea of a lot of fat may be quite different from yours. Ideas for increasing fat intake include:

Fried foods (use soy or canola oil for the best essential fatty acid profile):

peppers, mushrooms, onions added to tomato sauce

stir fry vegetables over low protein rice

Added fats (margarine, Miracle whip, butter, mayo)

Rich's Rich Whip or Cool Whip (NOT Lite)

Coffee rich (use on cereals, in soups, shakes, formula)

Canned frosting (eat right out of the can as a snack)

Regular salad dressings

Extra olive oil on salads in addition to dressings
Vegetable dips (low pro Ranch dressing is a favorite)
Chocolate or almond bark
Low pro candy bars
Oil added to the formula (is distasteful to some; others don't mind)

Sugars

"Free" Foods:

Koolade, Tang, fruit punches
Iced Tea with sugar
Soft drinks
Jam/jelly
Honey
Syrup
Sugar, brown or white
Hard candy

Some women object to eating sugary foods and need to be reassured that some of these foods are included in the diet to provide sufficient calories. Women with gestational diabetes should avoid these foods, however there is no evidence that dietary sucrose ingestion causes gestational diabetes.

Low Protein Foods Low protein products such as flours, breads, crackers, cookies and pastas offer much needed variety and energy to the MPKU diet. They are made from flour that been modified by removing gluten and other proteins, rendering the flour much like a cornstarch consistency. The greater the acceptance and use of these products, the greater the adherence to the diet and weight gain. It helps to identify a few low protein "staples" such as bread or pasta that can be used as a basis for meals.

Low protein foods are available by mail order in the United States from the following companies:

- Cambrooke Foods, Framingham MA
- Dietary Specialties, Randolph, NJ
- EnerG Foods, Seattle, WA
- Med Diet, South Minnetonka, MN
- SHS N. America, Gaithersburg, MD

These companies offer both ready-made products and flours for making you own bread, cookies, cakes, and snack foods. Recipes using these products are available from the companies or in low protein cookbooks such as Low Protein Cookery for PKU and Bread Machine Baking for PKU (both by Virginia Schuett).

These foods are very important for providing variety and calories in the diet, yet at first, are foreign in the way they cook, look and taste. In our experience, the women who eat low protein foods frequently gain weight the best and have the best metabolic control.

The key to success in using low protein foods is to work with a person who has experience using such products and can help identify simple, satisfying recipes that can be substituted for high protein foods. Too often, women start off the diet eating only fruits and vegetables. After a few weeks the diet becomes monotonous, the woman loses weight (driving up blood phenylalanine levels) and becomes discouraged about the lack of variety in the diet, which increases the likelihood of her not adhering strictly to phenylalanine limitations.

The Resource Daughter will usually need help determining which low protein foods to order and which low protein food companies have the best products and prices. Often, paying for the foods is a major obstacle. Many states now mandate that health insurance cover the cost of low protein foods; nevertheless, women often need help working through the insurance system to determine what they are entitled to and how to go about getting reimbursed.

Many of the special-order low protein foods are all considered "free" foods because they are so low in phenylalanine. Not all recipes in the PKU cookbooks are free, however. Look at the nutritional analysis to determine how much phenylalanine is in the serving size the woman will eat and help her figure out how much is allowed on her diet.