Image 1. Simplified PKU Diet; Department of Nutrition, Boston Children's Hospital

Simplified PKU Diet

Medical Food (Formula)

- · Medical food provides protein that is phenylalanine-free or low in phenylalanine.
- Medical food takes the place of high protein foods such as meat and dairy products

Food

- · The diet includes foods that are low in protein/phenylalanine.
- Count intake using the guidelines below.
- Recommended intake ___



Low Protein Foods - No Limit

Fruits

- All fruits that are fresh, frozen, or canned. Examples: apple, orange, pear, peach, melon, banana, pineapple, nectarine, berries, plum, kiwi, mango, grapefruit, clementine, fruit cocktail, applesauce, grapes
- Dried cranberries, raisins, prunes, and apples.







Vegetables

Most are low in protein - no limit Examples: asparagus, brussels sprouts, carrot, celery, cucumber, lettuce, onion, beet, cabbage, green pepper, red pepper, zucchini, cauliflower, squash, mushrooms, green beans, wax beans, artichokes, eggplant, tomato, plantain, raw spinach, raw arugula, and raw kale





Specialty Low Protein Foods

Most are low in protein* Examples: low protein pasta, rice, bread, bagel, cereal, broth, cookies, peanut butter substitute



*Discuss with dietitian about where to order, which to count

Other Foods

- Contain sugar and fat, but no protein, add extra calories
- Examples:
 - · Soft drinks such as lemonade, soda, iced tea (avoid aspartame)
 - · Fats butter, margarine, vegetable oil, mayonnaise, salad dressings (if label indicates zero protein)
 - Other nondairy creamer, coconut milk yogurt and ice cream; jelly, jam, sugar, maple syrup, popsicle, Italian ice, sorbet, marshmallow, hard candy







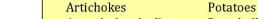






Medium Protein Foods - Need to Count

These vegetables/fruits are higher in protein and must be counted



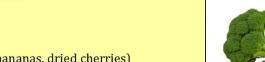
Arugula (cooked) Portabella Mushroom

Avocado Seaweed

Broccoli Sundried tomatoes
Collards (cooked) Sweet Potatoes
Corn Swiss Chard

Kale (cooked) Yam

Peas



Dried fruits (e.g. dried apricots, bananas, dried cherries)



Bread and Cereal Products

- Read food labels- look for the lowest in protein Examples:
 - Breakfast foods: toast, bagel, English muffin, waffle, pancake, cereal, small muffin, donut
 - Lunch and dinner foods: bread, pita, roll, tortilla, rice, pasta
 - Snack foods: potato chips, corn chips, rice cakes, Goldfish, crackers, popcorn, cookies





High Protein Foods (Avoid)

- · These foods are high in protein
 - Meat (All types-beef, lamb, pork, chicken, turkey, fish, etc.)
 - Food
 - Dairy products (cheese, yogurt, ice cream)
 - Nuts and seeds (including nut/seed butters)
 - Soy protein (tofu tempeh, many vegetarian burgers)
 - Legumes (kidney beans, garbanzos, lentils, split peas, hummus)







Boston Children's Hospital Metabolism Program

Fran Rohr, MS RD Ann Wessel, MS, RD Leslie Martell, MS, RD

Rev. 10/11/2016

Image 2. Explanation of Low Protein Diet; Metabolic Nutrition Program, Division of Medical Genetics, Icahn School of Medicine at Mount Sinai



Department of Genetics and Genomic Sciences Icahn Institute for Genomics and Multiscale Biology

METABOLIC NUTRITION PROGRAM

Explanation of Low Protein Diet

The low protein diet is divided into 4 categories:

- 1. Foods not allowed (completely eliminated from diet)
- 2. Foods allowed in limited quantities, called "limited items"
- 3. Foods allowed in unlimited quantities (as much as desired)
- 4. Special formula

1. Foods not allowed

- All Meat and meat products (includes gelatin and bouillon)
- Fish, fish products, and all types of seafood.
- Milk in any form, dairy products and all derivatives (such as eggs, cream, cheese, ice cream, sherbet, yogurt, etc.)
- Legumes such as soybeans, lentils, split peas, lima beans, green peas, any peas or beans (except green beans or wax beans), or any isolated protein derived from these sources such as vegetable protein, soy protein, soy flour, soy sauce, soybean curd (tofu), etc.
- Nuts and nut butters (including coconut and chestnuts)
- Seeds (all including sesame seed paste (Tahini).
- Any commercial product or condiments containing protein unless approved by the nutritionist.

2. Foods allowed in limited quantities

- Corn (including popcorn and corn chips)
- Potatoes (including French fries, potato chips, and yams)
- Rice and rice products
- Breads and baked goods
- Commercial breakfast cereals containing less than 2 grams of protein per cup
- Oranges, orange juice, bananas and plantains

The amount of all limited items and foods containing protein must be strictly controlled. Patients may be allowed 1 to 3 limited items or 0-10 grams of protein daily depending on doctor's or nutritionists orders. If blood levels cannot be maintained within a satisfactory range the number of limited items is reduced. Portions of all limited items and any food containing protein must be measured. For the correct potion sizes see Portion Sizes for Limited Items

3. Foods allowed in unlimited quantities

- All vegetables except the ones listed as limited or marked with an asterisk. See <u>Allowed</u>
 <u>Vegetables</u>
- All fruits except the ones listed as limited. See Allowed Fruits
- Special low protein products such as wheat starch bread, wheat starch, low protein baking mix, and all products made from these items. Sold as low protein foods from specialty companies.
- All condiments and foods which are free of any food or derivatives of foods which are unallowed or limited. See Allowed Condiments and Unlimited Foods
- All beverages which are free of any food or derivatives of foods which are un-allowed or limited. <u>See Allowed Beverages</u>

4. Special Formula or Amino Acid Supplement

The special products are prescribed by the metabolic nutritionist to supply adequate protein, calories, vitamins, and minerals for normal growth and development.

ALLOWED FRUITS

acerola nectarines

apples oranges (limited)

apricots passion fruit

bananas (limited)paw pawblackberriespapayascherriespeaches

cranberries pears
currants persimmons
dates pineapple

figs plantains (limited)
grapes (all varieties) plums (all varieties)
grape fruit

grapefruit pomegranate quava prunes

guava prunes kiwi fruit quinces "emu apple"

kumquat raisins
lemons raspberries
limes rhubarb
lychees strawberries

lychees strawberries loquat sapodillo mangos tangeloes melons (all varieties) tangerines

watermelon

NOTE: Limited fruits must be counted as limited items when large portions are eaten (see Portion Sizes for Limited Items), These fruits do not have to be counted as limited items, however, when used in small quantities such as in cooking (ex: banana bread), in fruit salad, etc.

ALLOWED VEGETABLES

Vegetables marked with an * when eaten in large quantities, add a significant amount of protein to the day's intake. Do not eat more than one vegetable marked with an asterisk per meal, and ½ cup max. They may be used freely in small quantities such as vegetable stews and mixed dishes. The number in parentheses is for protein content in ½ cup, cooked except for parsley.

acorn squash

anise

*artichoke (2.43 g)

asparagus

*bamboo shoots (0.92 g) basella (Indian spinach)

beets

beet greens
*broccoli (1.86 g)
broccoli rabe

*brussel sprouts (1.99 g)

burdock

butternut squash

cabbage carrots

*cassava = yuca, manioc (1.5g)

*cauliflower (1.14 g)

celery

celeriac (celery roots)

chard

chayote squash

chicory

Chinese cabbage (Bok choy)

chives

*collard greens (1.2 g) coriander (chinese parsley)

corn (limited) cucumber

dandelion greens

dasheen (Japanese taro) eggplant (aubergine)

endive escarole

*fennel (finocchio, 0.54g)

garlic ginger root

green beans (string beans)

hearts of palm

hubbard squash horseradish root *kale (1.24 g) kohlrabi

lettuce (all varieties) *lotus root (0.95 q)

*mushrooms (1.13gm, cooked)

mustard greens

okra olives onions

leek

*parsley (0.89 g)

parsnips

peppers (red and green)

potatoes (limited)

pumpkin purslane radishes rutabaga sauerkraut

*salsify (oyster plant, 1.84 g)

scallions shallots spinach, raw

spinach, cooked (2.6g) squash (all varieties) sorrel (dock, sourgrass)

sweet potato, yams (limited)

tampala leaves tomatoes

turnips

*turnip greens (0.82 g)

watercress water chestnuts wax beans

yautia (mélange, limited)

zucchini

ESTIMATED PORTION SIZES FOR LIMITED ITEMS

Containing 2 grams of protein

1 serving potato = 1 small potato (2 inch diameter)

1/2 large potato (4-3/4 inch X 2-1/2 inch)

1/2 cup mashed potatoes (no milk)

20 potato chips (2 inch diameter) or a 1 - ounce bag

10 thick French fries (1/2 X 1/2 X 2 inch)

20 very thin French fries (McDonald's type – small order) note: medium = 4 grams and large = 6 grams

1 serving corn = 1 small ear of corn on cob (3" long)

1/2 cup corn

1 1-ounce package Fritos corn chips

1-1/2 tablespoon un popped popcorn (110 kernels = 25.5 g)

1- 1/2 corn tortilla (6 inch diameter)

1 serving rice = 1/2 cup cooked brown or white rice

1 serving cereal = 1 cup allowed ready-to-eat breakfast cereal

1 cup cooked rice cereal

1 serving orange = 1-1/2 medium orange *

10 ounces orange juice1 cup orange segments

1 serving banana = 2 small bananas (7-3/4 inch X 1-1/4 inch)

3/4 cup mashed banana

1-1/4 large banana (9-3/4 inch X 1-1/2 inch)

*NOTE: Florida oranges contain less protein than California oranges, therefore serving sizes are based on Florida oranges.

These are **estimates**, please **read the food labels on packaging**.

ALLOWED CONDIMENTS/SAUCES

Jelly/Jam
Syrup (all types
Catsup (any brand)
Mustard (any brand)
A-1 Steak Sauce

Horseradish

Imitation butter flavoring

Flavoring extracts

Oil-based salad dressing Progresso pizza sauce

Buitoni marinara spaghetti sauce

Ragu pizza sauce

La Victoria green taco sauce

Jello pudding and pie filling (lemon, vanilla, butterscotch)

Royal pudding and pie filling (lemon, vanilla, butterscotch and coffee flavors)

Royal instant pudding and pie filling (lemon, vanilla, butterscotch and coffee flavors)

Tapioca

Relish

Margarine

Arrowroot

Baking powder

Herbs and spices

Ready-to-eat frosting

Old el Paso taco sauce

Tomato sauce, tomato paste

Hunt's prima salsa spaghetti sauce

Buitoni mushroom spaghetti sauce

Baking Soda

*If instructions call for 2 cups milk, substitute ¾ cup rich whip mixed with ¾ cup water.

Only royal instant pudding is allowed.

ALLOWED BEVERAGES

Tang (all flavors) V-8 cocktail vegetable juice

HiC fruit punches and fruit drinks Kook-Aid (all flavors)

Carbonated beverages (ex. Pepsi, Coke, Dr. Pepper, Gingerale, Sprite, etc.)

No beverages containing Aspartame are allowed.