

Nutrition and Huntington's Disease

A Guide for Families
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I. Introduction

More and more people are coming to realize the importance of good nutrition in maintaining health and preventing disease. Advice has been issued by many organizations about diet as it relates to the major killer diseases, heart attacks, strokes, and cancer. This advice focuses on reduction of fat and cholesterol, and increased consumption of fruits, vegetables, and grains. While this is sound advice for the majority of the population, there are some situations in which other factors take priority in the maintenance of optimal nutritional status.

People with Huntington's Disease may have much higher calorie needs than an average person, possibly due to chorea, metabolic changes, or a combination of both. There is also some anecdotal evidence that maintaining a body weight slightly above "desirable" weight will facilitate control of the disease. This would place an even greater emphasis on maintaining a high caloric intake. Of course, along with increased calories, people with HD need to maintain an adequate intake of all nutrients. They can easily meet the Required Daily Allowances (RDNS) for vitamins and minerals with a little planning, attention to a varied intake, and appropriate use of supplemental feedings.

The purpose of this booklet is to provide information about how to accomplish these goals. It is hoped that by making practical information about how to apply this dietary guidance accessible to patients and caregivers, the lives of people with HD can be enhanced.

II. Nutrition and Huntington's Disease

People with increased calorie needs must often be encouraged to eat even when they are not hungry in order to meet their nutritional requirements. This is usually not true of people with HD. They often have excellent appetites and sometimes eat very quickly. Frustration at being unable to get sufficient food down quickly enough without choking on it can exacerbate the psychosocial problems associated with food and eating in Huntington's Disease.

One approach to this problem is to provide six to eight smaller meals per day instead of the usual three. Frequent feeding can stave off the most high-level hunger and diminish eating urgency. Another approach is to offer frequent snacks or liquid supplements between meals.

As the disease progresses, and various activities of daily living become impossible, maintenance of independence in self-feeding becomes increasingly important. Foods should be selected with this capacity in mind. Items that are easier to manipulate may be the most appropriate. Also, with diminished capabilities in other areas, catering to food preferences provides an important psychological boost.

When it is no longer possible for the HD patient to meet his or her nutritional needs with an oral intake, feedings may be initiated. Tube feedings may be given either as a supplement to an oral diet, or as a sole means of nutritional support. This type of feeding is discussed further in Chapter 6 (see page 14).

III. Nutrition Needs in Early, Middle, and Advanced Stages of HD

While nutritional issues and eating difficulties may vary during the course of HD, maintaining quality of life and maximum functional ability is paramount throughout.

In the early stages, depression resulting from the diagnosis can cause decreased appetite and/ or changes in usual eating habits. Some people have reported food "cravings," especially for high carbohydrate foods. There is no harm in indulging these cravings, as long as other foods are eaten to maintain a balanced diet. A multi- vitamin supplement providing 100% of the RDA can offer some nutritional insurance. However, there is no evidence that excessive amounts of any nutrient are of benefit in HD.

Moving into middle stage HD, dysphagia (swallowing difficulties), choking, difficulty with self-feeding due to choreic (dance-like) movements, and additional difficulties due to psycho- social issues may appear. Other sections of this booklet offer some suggestions, and health professionals such as registered dietitians, speech- language pathologists, and occupational therapists can also be helpful. Many techniques and tools are available to help maintain independence in eating.

As the transition is made to advanced HD, dependence on others increases. Feeding another person is a loving and intimate activity that can be enjoyable or stressful depending on how it is approached by the individuals involved. It takes patience, and sometimes a sense of humor, to cope with this aspect of care. Social and cultural factors can also play a large role.

When it is impossible to get an adequate intake from an oral diet, tube feeding may be considered. It is possible to do this at home, given proper training. Tube feedings may be used to supplement an oral intake, or they may become the primary source of nutrition.

IV. The Eating Environment

A five-star restaurant is likely to be as well-known for its "ambience" as for its food. Where we eat and what is going on around us have a major influence on our eating experience. This is just as true of people with HD as anyone else. When eating problems arise, surroundings can be a crucial factor in helping to overcome them. It is important to keep distractions to a minimum so that the person with HD can concentrate on the task at hand - eating. Certain things may be more distracting to some individuals than to others, so experimentation may be needed to come up with the ideal arrangements for eating. Avoiding potentially stressful conversations, turning off the TV, having appropriate lighting and using proper positioning (when necessary) can all contribute to ease and enjoyment of eating. Needs will change with various stages of the disease, so flexibility is key.

The HD Kitchen

Having the right equipment can make preparing and serving meals for the person with HD much easier. Of course, what becomes "essential" is a matter of individual needs and preferences, as well as budget.

- A blender and/or food processor can be very helpful in preparing shakes, soups and sauces, as well as pureeing favorite foods when a very soft consistency is needed. If possible, get one that has more than one container, to allow for preparation of multiple foods quickly.
- A juicer can be useful for a person with dental problems, who cannot chew fruits and vegetables well. If swallowing is a problem, try adding a thickening powder, or mix the juice into a thicker shake or soup for a nutritional boost.
- A cappuccino maker may seem like a luxury, but switching from regular coffee to cappuccino made with whole milk and added cream (or vanilla ice cream!) can add some needed calories.
- A pastry cutter or potato masher can serve many purposes in mixing and mashing foods. These are also useful for adding 'extras' like butter or sour cream into a dish.
- Dishes with sides, also called "soup plates" or "pasta dishes," can make picking up food much easier and less messy. They are readily available in most department, housewares or medical supply stores.
- 'Sports' cups with a cover and straw attached can be helpful in preventing spills. Spoons or forks with larger handles can make picking up food easier. Rubber sleeves for utensils can be bought at a medical supply store.
- A heated "baby dish" or a warming tray can keep food warm throughout a meal for a slow eater.

V. Nutrition Needs in Early, Middle, and Advanced Stages of HD

Maintaining weight is important for people with HD. Keeping weight up is easier if you include high calorie foods as much as possible and maximize the calories in favorite dishes. Dig out grandma's old "fattening" recipes! The following are some high calorie food ideas to get started:

- Premium ice cream - choose plain flavors, not ones with "chunks" or "mix-ins" which could cause choking.
- Cream - plain, whipped, or sour cream - can be added as a supplement or topping.
- Butter - can be melted and mixed with many foods such as vegetables, mashed potatoes or yams, or hot cereal.
- Creamed soups, chowders, or bisques - mix in a blender or food processor to get the best consistency.
- Mayonnaise - add liberally to tuna, chick- en, or egg salads.
- Peanut butter, tahini, or other 'smooth" nut butters - to avoid choking think smooth - NOT chunky.
- Cheese sauce - as a fondue to dunk pieces of soft bread, or as a topping for vegetables or potatoes.
- Blue cheese or Ranch style salad dressing - as a dipping sauce for "chicken fingers."
- Refried beans - as a side dish or in a soft tortilla.
- Avocado - slices to dunk in salsa, or mashed as a side dish.

When you're Too Tired To Prepare Meals

- When you have the energy and are able to cook, make double the entire recipe or amount of your favorite foods. Freeze the leftovers in single servings, or whatever size you will need, to heat up and eat another day when you don't feel like cooking.
- Use convenience foods for quick and easy meals. Try frozen dinners, especially those with extra portions included, canned soups, stews, chili, spaghetti, or quick cooking macaroni & cheese. Instant breakfast drinks can be blended with milk, ice cream and fruit. Be sure to choose foods with the right consistency.
- Order out occasionally from a nearby restaurant. Save a file of menus from restaurants that deliver, and keep it near your telephone. Put one or more "stars" next to foods that you have tried before that were especially good.
- Keep lots of sandwich "fixings" on hand for a quick and easy meat or snack. Some high- calorie choices would be: bologna, liverwurst, salami, or most types of luncheon meat, and cheddar cheese, Swiss cheese, or most other types of hard cheese. Don't forget to include a spread such as mayonnaise or Russian dressing.
- Ask friends and family to bring food or to help you cook meals.
- See if any commercial meal service plans are available in your community. Such plans usually run by subscription, and will provide a package of reheatable foods for a specified number of meals per week. Some plans include delivery to your home.

VI. Tips for Coping with Swallowing Problems

(Also see *Coping with Speech and Swallowing Difficulties in Huntington's Disease* by Lynn Rhoades, MS, CCC-SLP, HDSA 1996.)

- Pay attention to which foods and/or beverages seem to cause coughing and choking. Identifying "problem foods" can save further trouble later on. Dry, "puffed," flaky or crumbly foods are often the culprits.
- Cut meat and vegetables into small pieces, and take small amounts in each bite.
- Choose soft, moist foods. Try mashed potatoes, noodles, soups, casseroles, yogurt, puddings, gelatins, soft cheeses, ice cream, milk shakes, creamed cereals, and macaroni and cheese.
- Add moisture to your foods with gravies, cream sauces, salad dressings, broth, sour cream, mayonnaise, or butter.
- Chew your food well. Sip liquids with a straw.
- Stay away from very acidic or spicy foods that may burn your throat if irritation is a problem. These include.- citrus fruits and their juices, tomatoes and tomato juice, and spices like red pepper, black pepper, chili powder, and hot curry powder.
- Try adding a thickener (like Thickit or Thicken-Up) or cornstarch to liquids to make them easier to swallow.

VII. Tube Feeding

Tube feedings are given when a person is unable to eat or tolerate enough food and/or oral supplements to meet his/her nutritional needs. Specific feeding instructions depend upon the amount and type of feeding to be given, and the type of tube being used. If you will be taking care of someone who is on tube feeding, you will need specific directions from your healthcare providers (dietitian, doctor, and/or nurse).

There are three basic kinds of feeding tubes:

1.) A naso-gastric tube (NGT) is threaded through a nostril, down the throat, and into the stomach. This type of feeding tube is usually temporary and does not require surgery to be put in. One disadvantage is that it can be pulled out accidentally. A naso-gastric tube also interferes with swallowing, which is a disadvantage if the person is also able to eat.

2.) A peri-epigastric tube (PEG), or gastrostomy tube (G-tube), is a tube that is implanted through the abdomen into the stomach. It functions in essentially the same way as an NGT. Formula flows through the tube into the stomach. It is usually for long-term use. As it does not pass down the throat, the patient can receive both tube feedings and an oral diet. It is a good idea to continue oral feeding for as long as possible, even if the quantities and choices are not nutritionally significant, as this can provide significant psychological benefits.

A new type of tube in this category is called a "button" tube. It is a very short tube attached to the stomach with a longer "snap on" tube for use during feedings. When the tube is not in use, a plastic cap covers the opening. This can be useful for people who do not receive a feeding constantly over 24 hours (e.g., bolus feeding, feedings run only at night, etc.).

3.) A jejunostomy tube (J-tube) is implanted below the stomach, directly into the small intestine. It functions similarly to tubes leading into the stomach, but with several differences. The advantage of a J-tube is that it reduces the risk that formula will back up the esophagus into the trachea and lungs. This is called "aspiration," and for someone at high risk of aspiration, the J-tube might be the preferred option. Increased probability of diarrhea, and increased probability of the (very narrow) tube getting clogged, are some disadvantages.

Feedings are usually either 'continuous' or "bolus" servings. As the name implies, continuous feedings run down the feeding tube at a fixed rate throughout the day and/or night. Tolerance problems are usually minimized when feedings are given in this way. A very weak or debilitated patient may need to be fed continuously in order to tolerate enough intake to meet his/her needs. Patients at risk for regurgitation because of limited stomach capacity need continuous feeding.

Most patients who are just starting out on tube, feedings are given continuous feedings and then gradually changed over to bolus feedings.

Bolus feedings are essentially the equivalent of a meal, consisting entirely of formula. A "bolus" is a set amount of formula run down the feeding tube at specific times during the day. These usually, but not always, correspond to breakfast, lunch, and dinner times. Some regimens include one or more bolus "snacks" as well, for a total of four to six feedings per day. Bolus feedings are usually more convenient for care-givers, since feedings are administered only at specific times, and larger amounts are given at each sitting.

It should be noted that for some people, the weight gain achieved after the gastric tube insertion leads to an improvement in swallowing ability. This improvement may allow the person to resume eating.

Some Additional Tips About Tube Feedings

1. Position the person so that he or she is sitting up, or at least so that the upper body is above the level of the stomach.
2. Practice good sanitation. Wash your hands before handling feeding equipment. Wash feeding bags with water. Do not use soap, as it will stick to the inside of the bag and get into the formula. This can cause diarrhea or other unpleasant consequences.
3. Feedings should be given at room temperature to minimize risk of cramping and/or diarrhea. Open cans of formula should be kept in the refrigerator, and discarded if not used within 24 hours. They should be taken out 15-20 minutes before a feeding and allowed to warm up to room temperature.
4. Always flush the feeding tube with water after a feeding. This will help to prevent the patient from getting dehydrated. It will also help to prevent the tube from getting clogged.
5. If medications are to be run down the feeding tube, always be sure they are finely crushed. Flush the tube with water to wash them down.
6. If the tube seems to be clogged and a small flush of water isn't effective, a flush of about 100cc of cola may do the trick. You can also try dissolving a quarter teaspoon of meat tenderizer in a teaspoon of water and placing it into the tube. Wait five minutes before flushing again. If none of these strategies work, contact your healthcare providers for advice.

VIII. Selected High-Calorie Recipes

BAKED CHICKEN SALAD

4 cups diced cooked chicken
1 1/2 cups cooked rice
1 can cream of chicken or cream of mushroom soup
1 cup mayonnaise
salt and pepper to taste 1 cup shredded cheese

Mix chicken, rice, soup, mayonnaise, salt and pepper together and place in a shallow baking pan. Top with shredded cheese. Bake at 350°F for about 30 minutes. Makes four servings. Each serving provides 794 calories.

Approximate exchanges are: 8 meat, 1 bread and 5 fat

HAMBURGER STROGANOFF

1 pound ground beef
1/2 teaspoon Worcestershire sauce
3 tablespoons flour
1 cup sour cream
1/4 cup butter
1 teaspoon salt
1/4 cup chopped onion 1/4 teaspoon pepper
1/2 pound fresh mushrooms, chopped

Melt butter in a large skillet and brown ground beef. Slowly mix in flour. Add chopped onion mushrooms and seasonings. Cover and let simmer for about five minutes or until mushroom and onions are soft. Add sour cream and mix well. Serve over soft noodles.

NOTE: This recipe also works very well in a crockpot (slow cooker). Brown the ground beef first. Then add all other ingredients and cook ,medium" heat for about 8-10 hours. Makes four servings. Each serving provides 581 calories

Approximate exchanges are: 5 meat, 1/2 bread and 7 fat

"IMPOSSIBLE" QUICHE

Butter a large pie pan and sprinkle with the following:

12 slices of bacon, fried and crumbled
1 cup shredded Swiss cheese
1/2 cup finely chopped onion

Place the following in a blender or food processor and blend until well mixed:
2 cups milk
1/2 cup Bisquick
4 eggs
1/4 teaspoon salt
1 1/4 teaspoon pepper

Pour blended mixture over the bacon, cheese and onion. Bake at 350°F for 50-55 minutes or until well set and golden brown. Allow quiche to cool for five minutes before serving.

NOTE: This recipe can be made in many variations depending on what you like and happen to have on hand. Shredded carrots, broccoli, spinach, mushrooms, shrimp, crab meat and different types of cheese can be substituted for the items listed above. Makes four servings. Each serving provides 597 calories.

Approximate exchanges are: 2 meat, 2 bread and 9 fat

APPLE KUCHEN

1 cup all-purpose flour
1 cup whole wheat flour
1/2 teaspoon salt
2/3 cup brown sugar
4 McIntosh apples, peeled, cored and quartered
1/2 teaspoon nutmeg
1 1/2 teaspoon cinnamon
1/2 cup chopped walnuts
1/3 cup butter
2 eggs, beaten
1 cup sour cream

Sift flours and salt into large bowl. Reserve two tablespoons brown sugar. Set aside. Add remainder of sugar to flour mixture and mix well. Add butter and walnuts and mix. Pack into 9- round cake pan. Arrange apple sections over mixture and sprinkle with two tablespoons brown sugar. Bake at 375°F for about 15 minutes or until apples are soft. While the above is baking, combine eggs, sour cream, cinnamon and nutmeg. Remove apple and crust mixture from oven and pour sour cream mixture evenly over the top. Sprinkle with a little more brown sugar. Return the kuchen to the oven and bake for an

additional 30-40 minutes. Serve warm or cooled. Makes four servings. Each serving provides 898 calories.

Approximate exchanges are: 1 meat, 3 bread, 12 fat and 1/2 fruit

BREAD PUDDING

5 slices of "day old" bread
3 cups milk, scalded
2 tablespoons soft butter
3 eggs
1/2 cup raisins
1/4 teaspoon salt
1/2 cup sugar
1/2 teaspoon vanilla
1/4 teaspoon cinnamon

Preheat oven to 350°F. Butter a medium sized baking dish. Toast bread lightly, then spread with butter while hot. Cut slices into quarters. Fit into prepared pan, overlapping slightly. Sprinkle raisins between slices, not over the top. Scald milk. Beat eggs slightly. Add salt to eggs, then all but two teaspoons of the sugar, then the hot milk and vanilla. Pour over toast and let stand 10 minutes, pressing toast down lightly once or twice to soak up milk mixture. Sprinkle cinnamon and remaining sugar over the top. Bake 25 to 30 minutes, or until a knife inserted in the center comes out clean. Serve warm or cold, plain or with whipped cream on top.

NOTE: For a finer texture, rip up the toast into very small pieces instead of cutting it into quarters. Makes four servings. Each serving provides 450 calories.

Approximate exchanges are: 1 meat, 1 bread, 3 fat, 1 milk and 1 fruit

CHEESE CAKE

6 8 oz. packages of cream cheese (3 lbs)
2 cups sugar
1 pint heavy cream
7 whole eggs
3 egg yolks
1 pint sour cream
2 teaspoons vanilla extract
1/2 teaspoon rum extract
1 1/2 cups crushed graham cracker crumbs
1/2 cup soft butter

Mix graham cracker crumbs and butter together and divide mixture in half. Press half of crumb mixture into the bottom of each of two 9- round cake pans. Cream together cream cheese, sugar, eggs and egg yolks. Add sour cream, heavy cream, and flavorings. Mix

well. Pour mixture into pans over the cracker crust. Bake at 350°F until set. Makes two cakes, eight servings each. Each serving provides 620 calories.

Approximate exchanges are: 1/2 meat, 13 fat and 1/2 bread

CHERRY CHEESE CAKE

18 vanilla wafer cookies
6 oz cream cheese
1/2 cup sugar
2 eggs
2 teaspoons vanilla
1 can cherry pie filling

Put paper cupcake liners into muffin tins. Place one vanilla wafer in each liner. Beat cream cheese, sugar, eggs and vanilla at medium speed. Pour into papers, on top of vanilla wafers. Bake 12-15 minutes in a 350°F oven. When cool, spoon cherry pie filling on top. Keep tarts in refrigerator until ready to serve. Tarts may also be garnished with whipped cream. Makes 18 servings. Each serving (1 tart) provides 133 calories.

Approximate exchanges are: 1/2 bread and 3 fat

PUMPKIN CHOCOLATE CHIP BREAD

2/3 cup butter
2/3 cup brown sugar
4 eggs
1 lb canned pumpkin
2/3 cup water
3 1/3 cup flour
2 teaspoons baking soda
1 1/2 teaspoons salt
1/2 teaspoon baking powder
1 teaspoon cinnamon
1 teaspoon ground cloves
1 teaspoon nutmeg
2 8 oz. packages semi-sweet chocolate chips

Preheat oven to 350°F. Grease two 9-x 5-x 3- loaf pans. Cream together butter and sugar. Add eggs, pumpkin and water. Add dry ingredients and mix well. Stir in chocolate chips. Pour into loaf pans. Bake 65-75 minutes. Makes two loaves, about six servings each. Each serving provides 586 calories.

Approximate exchanges are: 2 bread and 8 fat

ADDITIONAL RECIPES

Additional recipes which may be appropriate may be found in:

The Non-Cbew Cookbook

by *J. Randi Wilson*

Wilson Publishing Inc.

P.O. Box 2190

Glenwood Springs, CO 81602-2190

(800) 843-2409

\$23.95

The Dyspbagia Challenge

by *Pam Womack, RD*

NCES

1904 East 123rd Street Olathe, KS 66061-5886

(800) 445-5653

Catalog # 1760

\$11.00 plus shipping and handling

IX. Huntington's Disease Society of America

The Huntington's Disease Society of America is often the first place people go for information or assistance in coping with the effects of HD. HDSA can provide information about local resources, including knowledgeable physicians and other health professionals, genetic testing centers, support groups and long-term care facilities. Local social workers are available for information and support.

HDSA publishes and distributes a wide variety of informational materials, including newsletters, books, booklets, brochures and videotapes covering care, treatment, research and related topics.

The Society also funds over a million dollars in research every year through a grant and fellowship program. For more information, write to Huntington's Disease Society of America, 158 West 29th Street, 7th Floor, New York, NY 10001-5300, call (800) 345-HDSA or (212) 242-1968 or visit our site on the World Wide Web: www.hdsa.org