

## NUTRITION INTERVENTION IN HUNTINGTON'S DISEASE

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Severe weight loss is a chronic symptom of Huntington's disease (HD) even when caloric intake is adequate. The goals of nutrition intervention for Huntington's patients are to maintain weight and prevent aspiration. Benefits of weight maintenance are feelings of well-being, resistance to infection, and prevention of malnutrition.

Chorea (involuntary movements), a characteristic of HD, causes chewing and swallowing problems that interfere with food intake and may cause breathing of food into the lungs, leading to infection and even death.

Interventions to maintain weight and adjust food texture may be appropriate even from the initial diagnosis of HD.

Because weight is a measurable symptom of disease progression, it should be monitored and recorded regularly, on the same scale and under similar conditions each time by client or caregiver. Periodically, weights should be analyzed for trends up or down and the diet adjusted accordingly.

Weight loss may appear to be spontaneous or related to other physiological and/or psychological conditions. Weight loss may be aggravated by one or more of the following factors:

- Dysphagia (difficulty swallowing) which limits food intake;
- Increased energy expenditure due to chorea (involuntary movements) or to dystonia (rigidity);
- Change of psychological condition which limits food intake.

Dysphagia results from a decline in motor co-ordination associated with the basic chorea. Difficulties in chewing impede swallowing. The intricate swallowing process itself may become disordered, resulting in possible choking or aspirating, precipitating pneumonia.

Dry mouth from medications may compound chewing problems because reduced saliva may cause tooth decay and loss, which both also negatively impact chewing.

Appetite decline, due to physiological and psychological changes, also contributes to compromised caloric intake. The patient's progressive physical decline is associated with loss of appetite due to taste changes, depression, reluctance to eat for fear of choking (silent fear of

aspiration), feelings of fullness due to constipation, and nausea or dry mouth from medications. These are problems that the person with HD may be unable to identify or communicate, which leads to the impression of unreasonable refusal to eat. Misunderstandings on the part of a caregiver, of the client's refusal to eat, result in conflict, hostility, frustration, and despair. Depression heightens nutritional risk and nutritional risk heightens depression.

The diet should consist of balanced nutrition: adequate calories distributed among all food groups. Carbohydrates should be chosen from complex, not simple sugar, sources such as winter squashes, beans, and root vegetables. These will provide more lasting satiety and their fiber will aid bowel regularity.

**Fats**, the storehouse of energy in the diet, should be from heart-healthy VEGETABLE fats: unsaturated oils, such as corn, olive, canola that are liquid at room temperature. Vegetables carry no cholesterol and their oils do not contribute to the build-up of artery-clogging plaque that leads to heart disease and high blood pressure. Vegetable oils additionally provide satiety, flavor, and mouth feel. With double the number of calories of carbohydrates, fats can be liberally combined with carbs in soups, purees, casseroles, sautés, and stirfrys.

**Protein** (dairy, meat, poultry, fish), which metabolizes more slowly, provides energy over a longer period of time. It also builds muscle mass and strength.

The need to provide high calories conveniently, at reasonable cost, and within the four basic food groups, while respecting lifelong food preferences, demands a dietitian's experienced counsel. Because of the exaggerated number of calories required to maintain weight, often as high as 3,500 to 5,000 a day, HD clients need either high volumes of food or portion-controlled, highly fortified foods.

Preferred foods for fortification are the patient's lifelong favorites and culturally familiar "comfort foods." These can be combined with bland, high-calorie food fortifiers from all food groups. Commercially formulated supplements can also augment calories in the diet. No vitamin or mineral therapy has been found to make a positive difference in HD.

Calories can be added at meals, through frequent snacks between meals, and at night since HD clients are often awake for long periods. Menus that encourage intake through variety in flavor, temperature, and use of color, as well as a quiet atmosphere at meals, are conducive to increased consumption.

Altered texture foods (ground, chopped, pureed solids and thickened fluids) help to overcome chewing and swallowing problems. The commercial thickeners for liquids themselves carry calories. Since many consider altered texture "baby food," it is more effective to offer foods that have a soft consistency naturally, like yogurt, or sandwiches such as egg salad on soft bread.

Hydration is included in the weight maintenance program: fluids carry calories and prevent

constipation and the feeling of fullness, which is an appetite diminisher. Related to bowel regularity, high-fiber foods or use of commercial fiber supplements in food also contribute to a tonic feeling.

Medications may affect nutrition. The side effects on appetite or of changing dosage, as well as the need to give meds on an empty or full stomach, will all influence food intake.

Diet for other medical conditions will need to be taken into account. Mediating blood sugar and weight, cholesterol and sodium intake, gluten or lactose intolerance, are conditions that may moderate the weight goals.

Self-feeding problems arise from chorea or dystonia, both of which make clients unable to handle utensils to transport food from plate to mouth. Adaptive equipment, such as high-sided plates and noney cut-out cups, is available in the event of feeding problems. Clients may need to be fed. For special techniques, a Speech and Language Pathologist who specializes in dysphagia therapy can be consulted.

When weight cannot be maintained safely through food intake, it is the dietitian's function to alert the caregiving team that a feeding tube may need to be placed. This is both a team problem and a client-centered one. Decision-making can be facilitated by an Advance Directive that the client has prepared earlier in the course of disease and by the Health Care Proxy, if the client is incapable of making a reasoned decision.

In summary, HD clients may need:

- high calories, as much as 3500-5000 each day;
- altered-texture diets;
- assisted or total feeding;
- high fiber and hydration for bowel regularity;
- techniques to overcome the side effects of medications such as dry mouth and loss of appetite.
- people with HD need integration of their nutrition needs with their chewing and swallowing tolerances.