

## **Coping with Speech and Swallowing Difficulties in Huntington's Disease**

### *Disclaimer*

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### **Introduction**

A speech-language pathologist (SLP) can have a profound effect on quality of life for a person with Huntington's Disease (HD). I realized this one evening several years ago when, after an in-service presentation to family members, a young woman approached me and said, "I never knew that there were so many options available to my mother if her swallowing failed. I always thought she would automatically have to get a G-tube. It is such a relief to know that you can try all these other things...Thank you."

That woman's statement sums up the purpose of this booklet, which is to provide families with information that will hopefully remove some of the struggle and fear associated with living with HD. This publication will focus on common communication and swallowing problems that occur in persons with HD, what you can do at home to help manage these problems, and how an SLP, as well as other professionals, can be of service to you and your family.

### **Why Communications and Swallowing Symptoms Arise in HD**

As brain cells become depleted in Huntington's Disease, problems may develop in the following three areas: motor control (movement); cognition (thinking); and behavior. Speech and swallowing problems arise when the centers of motor or cognitive control are affected, causing muscle weakness or discoordination, chorea, and problems with memory, sequencing, new learning ability and problem solving.

Typically, speech and language functions are primarily controlled in the left side of the brainstem (at the base of the brain). Cognitive function is believed to be controlled in the right side of the brain. HD typically begins in the caudate nucleus and putamen, which are located in the central part of the brain (core), and spreads to these other control centers, causing communication and swallowing problems.

## **Communication**

Problems with communication and cognition vary in nature and severity from person to person. Although there are commonalities, no two people with HD are exactly alike, and as the disease progresses, the variability continues. The following list summarizes problems that people with HD may experience the same areas of difficulty throughout the course of the disease, with severity varying from stage to stage.

### *Communication problems*

- o Dysarthria (muscular weakness, slowness, or incoordination of the lips, tongue, throat, and jaw)
- o Apraxia (disruption in programming and sequencing muscle movements for speech)
- o Diminished rate control (talking too fast or too slowly)
- o Poor voice quality (horse/harsh, breathy, volume too low or too high)
- o Discoordination of breathing and voice
- o Word finding difficulties
- o Short length of utterance (person only responds with one or two words)
- o Misarticulations (incorrect pronunciation of sounds)
- o Lack of initiation (inability to initiate conversation)
- o Perseveration (person gets "stuck" on certain words or phrases, repeating them often and at inappropriate times)
- o Echolalia (person keeps repeating back what you've just said)
- o Difficulty with monitoring pragmatic skills (turn-taking in conversation; reduced ability to maintain a topic or switch topics appropriately)

### *Inability to speak*

- o Stuttering (difficulty beginning a word or sentence, with repetition of sounds)
- o Difficulty understanding information
- o Difficulty reading and writing

### *Cognitive problems*

- o Diminished memory, immediate and short-term (long-term memory usually remains intact\*)
- o Poor reasoning/judgment
- o Reduced problem-solving ability
- o Difficulty sequencing/organizing ideas
- o Concentration problems/distractibility/short attention span
- o New learning ability diminished
- o Problems with numbers and mathematic computations

NOTE: It should be emphasized the most people with HD are receptively intact, and that even during the end stages of the disease, they can understand what is being said to them.

## **How the speech-language pathologist can help**

The speech language pathologist can be helpful at all stages of the disease. In the early stages, he/she can assist with problem solving and developing strategies to help persons with HD compensate for some of the problems they might be experiencing. As the disease progresses, the role of the SLP evolves into helping preserve and maintain the person's highest level of communication and swallowing. Early intervention and involvement with therapeutic professionals is best because people can learn compensatory strategies more successfully during the early stages of HD and can then apply them throughout the course of the disease.

The SLP can also evaluate a person's ability to use augmentative or alternative communication devices and techniques, which can be as simple as a word/picture board, or more complex, such as an electronic device that speaks for the person. After determining a person's level of ability for using such techniques, the SLP begins to focus on personalizing the technique or method of communication.

For example, the SLP might work with a person with HD and his or her family to create a word/picture board tailored to the person's environment (whether it be a nursing home or private residence) or flexible enough to be carried around. Where an electronic device might be beneficial, the interest and motivation of the person with HD to use it should be evaluated. If there is evident motivation, then the device should be made easily accessible.

#### *Suggestions for improving communication*

Although alternative methods of communication are available, people with HD generally prefer to attempt verbal communication for as long as possible, even if their speech becomes hard to understand. The SLP and family members can often help by encouraging the speaker to:

- o Speak more slowly
- o Say one word at a time
- o Repeat the word or sentence when necessary
- o Rephrase the sentence
- o Exaggerate the sounds
- o Speak louder (take deep breath before speaking)
- o Describe what he/she is trying to say if he/she can't think of the word
- o Indicate the first letter of the word
- o Use gestures Keep sentences short
- o As a supplement to speech, use alternative communication techniques such as word boards, alphabet boards, picture boards, electronic devices

Some suggestions for the listener are:

- o Eliminate distractions (TV's, radios, large groups of people)
- o Keep questions/statements simple
- o Ask one question at a time
- o Use yes/no question format as much as possible
- o Pay attention to gestures and facial expressions/changes
- o If you do not understand what is being said, don't pretend that you do. Ask for clarification, or repeat what you think was said in the form of a question, such as, "Did you say...?"
- o Try to keep to familiar topics
- o Encourage the speaker to use his/her specific compensatory strategies
- o Allow enough time for the person to convey his/her message
- o Most importantly, be patient with the speaker

People with HD should seek out an SLP at the point when communication with others becomes frustrating and upsetting, or when the areas of memory, thought processing and organization of thought become problematic.

There are also compensatory strategies for cognitive problems that can be implemented in the home. Some examples are:

#### *Problem*

Poor orientation to time and place

#### *Strategies*

- o Keep a large calendar visible
- o Display a large, visible clock
- o Post signs on walls stating location

#### *Problem*

Diminished memory

#### *Strategies*

- o Post a schedule of daily routine
- o Establish routines for all activities, e.g., place keys or glasses in same place daily

- o Label cabinets and drawers
- o Keep a memory log book - like a diary - in which the person with HD can write down and refer back to what he/she has done, with whom, and when.
- o Keep an appointment book for social events, doctor's visits, and other dates and occasions to be remembered

#### *Problem*

Reduced problem-solving ability

#### *Strategies*

- o The person with HD should discuss with a family member possible problems that could occur in the home. Solutions or steps to be followed should be anticipated, written down and kept in an obvious place
- o Follow these set guidelines for problem solving

These are just a few examples of how a person experiencing cognitive problems can make life easier at home. An SLP can provide assessment, guidance, and further suggestions on the use of compensatory strategies.

#### *Swallowing*

Swallowing problems (also known as dysphagia) are common among people with HD. Statistics have repeatedly shown that the number one cause of death among persons with HD is aspiration pneumonia. This can occur when food or liquid enters the airway rather than the esophagus during eating or drinking, and then forms a collection in the lung that can become a pneumonia.

The involvement of an SLP can be helpful at all stages of the disease in maintaining the highest possible level of swallowing function. This intervention will help to prolong the quality of life and may lower the risk of death caused by pneumonia.

There are many steps that can be taken to preserve safe swallowing for as long as possible. First, let's look at the problems that people with HD frequently encounter:

#### *Swallowing Problems Associated with HD*

- o Impulsivity or difficulty controlling amount of food or liquid intake
- o Difficulty controlling rate of food or liquid intake
- o Difficulty chewing or manipulating food
- o Delayed swallow reflex (doesn't kick in even when food moves to the back of the throat)
- o Holding food/liquid in mouth
- o Difficulty initiating a swallow at all
- o Inability to swallow
- o Incomplete swallows where food or liquid is left in the mouth and/or throat
- o Lack of coordination between swallowing process and breathing or speaking
- o The need to swallow repeatedly for each bite/sip
- o Chorea of the oral or pharyngeal muscles (tongue, lips, jaw, throat, esophagus)
- o Drooling and/or spillage of food or liquid from mouth

The SLP can evaluate a person's swallowing function and make recommendations that involve positioning issues, feeding techniques, diet consistency changes, and education of the person with HD, family members, or caregivers. Special testing known as videofluoroscopy (or a Modified Barium Swallow) can be done by a speech-language pathologist and a radiologist to determine if a person is actually aspirating a particular consistency. This test provides an inside view of a person in the act of swallowing food or liquid and can be a useful tool in developing strategies for safe swallowing.

#### *Warning Signs*

The following signs at mealtime may indicate swallowing problems:

- o Coughing
- o Choking
- o Gurgly voice quality
- o Wet sounding breathing
- o Spillage of food or liquid from the mouth
- o Frequent throat clearing
- o Progressively slower rate of food intake
- o Regurgitation of food after it has been swallowed\*
- o Food or liquid left in the mouth after swallow
- o Difficulty manipulating food or liquid in the mouth
- o Frequent congestion\*
- o Frequent temperatures\*
- o Consistent or significant weight loss\*

Signs marked \* could be indicative of a serious and possibly unrelated medical condition and should be monitored by a physician. In general, if a person with HD experiences any one or a combination of the above problems, he/she should contact a physician and seek out an SLP for evaluation.

## SAMPLE CASES

### CASE I

#### *Symptoms*

1. Person was coughing and throat-clearing frequently when drinking thin liquids such as juice or coffee.
2. He was observed to drink all liquids from a cup and to throw his head back as the cup emptied.
3. Then he would cough hard.

#### *Problems observed during evaluation*

1. Suspected upper airway penetration (liquid entering the airway instead of esophagus).
2. Rate of intake too fast.
3. Size of sips too big.

#### *Solutions*

1. Person was given a straw. He took liquids with the straw and his coughing and wet vocal quality stopped. He now drinks all liquids, including coffee, via straw.
2. This keeps his head in a more forward position and helps to control the rate of intake (he tended to take big gulps with the cup).
3. He is given verbal reminders to take small sips.
4. Staff was educated about these strategies for safe swallowing.

### CASE II

#### *Symptoms*

1. Person holding food in mouth for long periods of time.
2. Swallows were delayed and food was left in mouth after the swallow.

#### *Problems observed during evaluation*

1. Chewing skills were observed to be ineffective.
2. Mouthfuls were too big.

#### *Solutions*

1. Person was placed on a finely chopped diet (regular consistency too much for her to chew).
2. Cues were given to alternate sips with bites and to place less food on the utensil.

### CASE III

#### *Symptoms*

1. Person feeding self and experiencing a lot of spillage of food/liquids from the mouth.
2. Significant weight loss.

#### *Problems observed during evaluation*

1. Severe dysarthria (muscles of mouth and throat weak).

2. Person holding head down when trying to swallow (leaning far forward).
3. Person now unable to chew.
4. Person not coordinated for self-feeding anymore.

#### *Solutions*

1. Person placed on pureed diet.
2. A slightly reclined position in bed for mealtimes.
3. Feeding now done by staff
4. Family allowed to bring soft favorites (candy, snack cakes, shakes).

#### *Strategies for Safe Swallowing in the Home of Care Facility*

1. Provide small bites and sips.
  2. Alternate bites with sips to help wash down the food.
  3. Make sure that the person is sitting as upright as possible; place pillow behind head/neck to prevent head tilting backwards.
  4. Control rate of intake, allowing enough time for previous bite/sip to be cleared. Look for rise and fall of larynx (Adam's apple) or lightly place two fingers on the person's Adam's apple to feel this rise and fall, which can signal the completion of a swallow.
  5. Person should swallow twice for every bite or sip to clear residuals from the mouth.
  6. Use gravies, sauces and condiments with dry foods or solids whenever possible.
  7. Crush medications in applesauce if a person is on a pureed (blenderized) diet.
  8. Avoid distractions during oral intake:
    - No TV
    - No talking with food in mouth
    - No large crowds
- o If the person is coughing a lot or showing other signs of swallowing problems, begin giving thicker liquids such as nectars, shakes, and tomato juice. Also give blenderized food until person can be seen by the physician and/or speech language pathologist. Further recommendations would follow a complete assessment.
  - o Produce a strong cough after each swallow (if voice sounds 'gurgly' or 'wet').
  - o Limit quantity of food placed on plate or liquid in cup at one time.

#### *Alternative Methods of Feeding (Nasogastric Tube, Gastrostomy Tube)*

As we have seen, there are many interventions and strategies that can help to preserve safe swallowing - some very simple and easily implemented at home. Others require evaluation and guidance from an MD or SLP. If all attempts fail, only then will it be necessary to discuss alternative methods of feeding.

Once this point has been reached, the task of deciding what is best can be difficult and emotional. This issue typically arises when a person with HD is in the end stages of the disease, although it may need to be addressed earlier in some cases. By the late stages of HD, the person is often unable to meet nutritional needs, tends to become dehydrated, or aspirates all possible consistencies - often reflected by frequent bouts of pneumonia. The eating process may have become unpleasant.

The following is a list of possible considerations involved in deciding whether or not to proceed with G-tube placement:

- o Are the person's wishes known or documented?
- o Is he/she dehydrated?
- o Is there severe weight loss?
- o Is the person suffering from malnutrition, with possible subsequent skin breakdown?
- o Does the person have frequent temperatures?
- o Does he/she have frequent bouts of pneumonia (especially aspiration pneumonia)?
- o Is there decreased, or lack of, enjoyment at mealtime?
- o Is the person unable to swallow without coughing/choking?
- o Does he/she have difficulty with respiration following oral intake?
- o Are any other interventions/strategies available?

o How important is eating to the person? Is eating a rewarding experience?  
The risk/benefit ratio of the enjoyment which eating brings versus the dangers of eating for this person should be carefully weighed.

A nasogastric tube is considered temporary and inserted through the nose. It goes down the back of the throat and into the stomach. A G-tube is considered more permanent but may be reversed. Placement requires a minor surgical procedure with insertion directly into the stomach. Shake like feedings and medications are introduced via tube.

In both cases, if a person is able, he/she may continue eating and drinking while the tube feeding provides the consistent caloric intake needed for weight maintenance, or gain, if so desired.

The issue of alternative feeding is difficult because it may extend a person's life span and thus prolong the disease. At the same time, families find it difficult to decide against alternative feeding because the end may be hastened by their decision. There are no easy answers and it can be very hard to gauge a person's quality of life at the time when this issue arises.

Ideally, family members should know in advance what the individual's wishes are with regard to G-tube placement (see Planning Ahead). It is important to discuss the issue beforehand as, when the time comes to implement the decision, the person may be unable to communicate his/her wishes or to provide a reliable yes/no response to questions regarding the issue.

At this juncture, the role of support personnel is critical. Professionals who can offer guidance and assist with this kind of decision-making include the:

- o Physician/Medical Director
- o Social Worker
- o Speech-Language Pathologist
- o Chaplain
- o Dietician
- o Biomedical Ethics Committee
- o Nurse

These professionals can answer questions regarding the ramifications of placement, or non-placement, of a tube. "What will happen if the tube is not placed and the person can no longer be fed?" "What will happen when we continue to feed someone who we suspect is aspirating frequently?" "What are the benefits of tube placement, and what are the possible complications?" "Does the person having the tube placement feel any pain? Does the person who does not get a tube and doesn't eat anymore feel any pain?"

These are common questions asked by families and answered as completely and sincerely as possible by the professionals.

When grappling with this decision, support can come from other areas such as clergy, or what is known as biomedical ethics, or bioethics, committee. This group of professionals, which can include physicians, nurses and patient advocates, may be requested to assist with decision-making on the issue of alternative feeding. Often, the committee makes decisions for a patient who is hospitalized and has no family involvement or other decision-makers to act on his/her behalf, or if there is a conflict between the person's wishes and family concerns. The best scenario is when a document can be produced that clearly indicates the person's wishes.

## **Planning Ahead**

It is tough enough to face a diagnosis of HD and to cope with early symptoms without having to look ahead, but the sooner these issues are discussed, the more control a person with HD can exercise over these crucial decisions.

People in the early stages of HD and those at risk should complete a "Living Will" or "Health Care Proxy." The Living Will is a clear statement of a person's wishes in terms of medical treatment and life-support should they ever become unable to communicate or understand information. This includes placement of a G-tube or nasogastric tube, as they are considered life-support. The Health Care Proxy enables a person to designate a third party to make these kinds of decisions on the person's behalf in lieu of a written request, or to insure that a written request is carried out.

Again, documentation and communication of these decisions are of paramount importance. It is crucial that these processes and communication are not impeded by HD.

## **Conclusion**

Huntington's Disease is a devastating illness that affects every aspect of a person's life. Although no treatment yet exists to stop or reverse the course of HD, health professionals of different disciplines can help to maximize quality of life for as long as possible.

In addition to speech-language pathologists, physical and occupational therapists can bring strategies and equipment to bear to make dealing with HD a little easier. Social workers, recreational therapists, dieticians, and others can all make important contributions.

Most importantly, the sooner the person with HD arrives for evaluation, the sooner the professional can implement a plan that will prolong independence and maintain the highest level of function for the longest period of time.

## **Resources**

How to Locate a Speech-Language Pathologist

1. Physician referral.
2. Phone book.
3. Call any major hospital. They should have a rehabilitation department that includes speech-language pathology.
4. Call the Speech-Language-Hearing Association (ASHA) at the following address or phone number:

American Speech-Language-Hearing Association  
10801 Rockville Pike  
Rockville, MD 20852 (800) 638-8255

## **For Further Information**

Additional publications about nutrition, communication, swallowing and related topics may be obtained from the Huntington's Disease Society of America, 505 Eighth Avenue, Suite 902, New York, NY 10018, (800) 345-HDSA, (212) 242-1968.

Visit our site on the World Wide Web: <http://www.hdsa.org/>. The following videotape is also available from HDSA, 505 Eighth Avenue, Suite 902, New York, NY or call (800) 345-HDSA (4372) for \$10 including shipping and handling: "The Gift of Caring: A Practical Guide to Managing Feeding and Swallowing Difficulties Associated with Huntington's Disease".

"The Non-Chew Cookbook, by J. Randi Wilson, 1985, 188 pages, which

offers easy recipes complete with caloric intakes, can be purchased for \$23.95 from Wilson Publishing Inc., P.O. Box 2190, Glenwood Springs, CO, 81602-2190, (800) 843-2409.

"Tomorrow's Choices: Preparing Now for Future Legal, Financial and Health Care Decisions" is a free publication available from the American Association of Retired Persons (AARP), 601 E. Street, NW, Washington, DC 20049. Ask for publication #D13479.

## **Glossary**

(Speech-related terms are defined in communication section)

**Alternative/Augmentative Communication**

Assistive devices to aid in communication, such as letter or word boards, picture boards, electronic devices, gestural systems.

**Alternative Feeding**

Nutritional intake provided by something other than oral feeding.

**Cognitive Skills**

Areas of thought processing, including problem solving, reasoning, judgment, sequencing, attention, memory, understanding of more complex information, concentration, and new learning ability.

**Compensatory Strategy**

A technique that can help make up, or compensate, for a particular loss of function or problem.

**Dysphagia**

The disorder of swallowing that can be characterized by problems within the oral cavity or the pharynx.

**Modified Barium Swallow**

Swallowing test conducted in the radiology department in which barium is mixed with different food consistencies and presented under videofluoroscopy (X-ray).

**Pharynx**

Throat.

**SLP**

Speech-language Pathologist