

HDSA Family Guide Series



Speech-Language and Swallowing Difficulties



Huntington's Disease
Society of America

Speech-Language and Swallowing Difficulties in HD

Family Guide Series

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Contents

Introduction:	4
What is a Speech-Language Pathologist (SLP)?	5
Speech Characteristics Associated with Huntington's Disease	7
• What makes speech acceptable and understandable to a listener?	7
• Mild impairment in speech/ language and cognitive function	8
• Moderate impairment in speech/ language and cognitive function	9
• Severe impairment in speech/ language and cognitive function	11
Swallowing Problems Associated with Huntington's Disease	12
How is Swallowing Assessed?	14
Swallowing Strategies to Minimize the Risk of Aspiration	16
• Diet modification	16
• Positioning	16
• Compensatory strategies	16
Dysphagia Management Options	17
• Continue to eat by mouth	17
• Tube feeding	17
Sample Cases	19
• Case I	19
• Case II	20
• Case III	21
Conclusion	22

Introduction

If you are reading this, you may already know that Huntington's disease (HD) can have a devastating impact on a person's ability to speak and swallow. It is an inherited autosomal dominant disorder, which means that each child of a parent with the expanded HD gene has a 50% chance of inheritance. The disease causes a progressive breakdown of nerve cells in the brain that result in a triad of motor, thinking, and behavioral disturbances. Speech and swallowing problems can arise due to involuntary muscle movement (chorea), weakness of the throat muscles, and cognitive impairment. The symptoms that an individual experiences, and the way these symptoms impact function, vary from person to person, and are also affected by the stage of the disease process.

This pamphlet has been developed to provide you with a basic understanding on communication and swallowing problems associated with Huntington's disease. The publication will focus on the most common communication and swallowing difficulties that occur in the different stages of Huntington's disease, and how the Speech-Language Pathologist (SLP), and other health care professionals, can help you and your family cope and manage these difficulties in order to improve or maintain the highest quality of life. Several cases are provided at the end of the publication to illustrate solutions to potential problems.

What is a Speech-Language Pathologist (SLP)?

A Speech-Language Pathologist is a person who specializes in the assessment and treatment of speech (dysarthria and apraxia) and language disorders (aphasia), impaired cognitive aspects of communication (e.g., attention, memory, problem solving, executive functions), and swallowing disorders (dysphagia) and is often involved in the care of a person with Huntington's disease.

- **Aphasia** - Difficulty saying or writing what you mean and/or making sense of words spoken or written
- **Apraxia** - Trouble sequencing the sounds in syllables and words
- **Cognitive** - language impairment – Difficulty with attention, memory, problem solving, and planning
- **Dysarthria** - Slow, weak, imprecise and/or uncoordinated speech due to muscle weakness and/or incoordination of speech muscles
- **Dysphagia** - Difficulty swallowing

Although helpful, it is not necessary for a SLP to be familiar with HD because these types of problems also occur in conjunction with other conditions.

When a person is first diagnosed with HD, it is beneficial to see a SLP for early intervention. At that time, the SLP will complete an initial evaluation in order to get baseline information on the person's speech and swallow function. The SLP will

also be able to give the person with HD valuable information about maintaining safe swallowing and different strategies to enhance communication. Baseline information also allows the SLP to develop a plan to educate the person and the family about potential interventions to help the person maintain independence as HD progresses. You can find a SLP by asking your physician for a referral or calling the American Speech-Language-Hearing Association (ASHA) at **(800) 638-8255**. The ASHA website also has a search engine where you can search for SLPs by state, at [asha.org/proserv/](https://www.asha.org/proserv/).

Speech Characteristics Associated with Huntington's Disease

What makes speech acceptable and understandable to a listener?

One can think of speech as being made up of many different aspects including: pitch, loudness, voice quality, articulation, respiration, resonance, and prosody.

- **Pitch** - The degree of voice highness or lowness
- **Loudness** - How well one can project his or her voice
- **Voice quality** - How well one can hold their pitch and how clear the voice sounds
- **Articulation** - The ability to pronounce sounds
- **Respiration** - Coordination of speech with breathing
- **Resonance** - The quality of the voice that is determined by the balance of sound vibration in the mouth, nose, and throat during speech
- **Prosody** - The rhythm, stress, and intonation during speaking that helps give meaning to a message

The speech disorder most associated with HD is hyperkinetic dysarthria, which is characterized by excessive, irrelevant, and involuntary movement of the muscles that a person uses to speak. The underlying movement disorder can affect every aspect of speech resulting in different perceptual characteristics. For example, speech can sound harsh or strained, hypernasal (overly nasal), breathy, or have variation in loudness levels. Speech can also lack prosody, which means that it lacks rhythm and intonation and sounds flat, with no emphasis. When one or more aspects of speech are impaired the result is “abnormal speech,” which can cause a breakdown in speech intelligibility.

Mild Impairment in Speech/Language and Cognitive Function

Changes in speech patterns that are noticed by both the speaker and the listener, but do not interfere with speech intelligibility are considered to be a mild impairment. At this stage, people are typically not referred for speech/language services because they are able to routinely verbalize and communicate without difficulty. However, if these changes interfere with the person's ability to participate in activities, if a person starts avoiding certain situations or activities because of their speech disorder, or if the person is developing abnormal behaviors to compensate, even in very mild cases, speech therapy may be appropriate. There is some benefit to seeing a speech pathologist early on in the disease process, before the speech disorder progresses to the point that it impacts speech intelligibility, especially if the person is using maladaptive behaviors in an attempt to compensate for the speech disturbance.

Some of the more common aspects of speech that are affected in the early stages of the disease include speech rate and voice quality. Often, a person with HD may speak at a rapid rate in an obvious attempt to get an utterance spoken before a movement disturbance occurs or the person's air runs out. Educating the speaker on the importance of maintaining an appropriate rate of speech and training appropriate breathing patterns during speech may improve overall speech intelligibility. Phonatory stenosis, a pattern found in some persons during early-stage HD, is a cluster of voice symptoms caused by brief, random closing of the vocal folds that may result in a perceptually harsh voice or strangled voice. Treatment for phonatory stenosis targets reducing tone in the muscles used for voice and may also include relaxation techniques.

Moderate Impairment in Speech/Language and Cognitive Function

Moderate impairment is defined as change in speech patterns that compromise intelligibility that are noticed by both the speaker and the listener. Just as in the case of the person with mild dysarthria, a person with moderate dysarthria may benefit from a speech evaluation and treatment. During this stage of the disease, abnormal respiratory, or breathing, and speech patterns may be evident. Abnormal respiratory behaviors include speaking on inhalation rather than exhalation or not starting the voice at the beginning of a breath. These abnormal respiratory-speech patterns will often impact other aspects of speech including stress patterning, rate of speech, and prosody (rhythm of speech). An adequate level of speech intelligibility may be achieved by teaching the person relaxation techniques, compensatory strategies that improve respiratory support, as well as respiratory and speech coordination.

At this stage, cognitive changes are likely to occur as well. The person may no longer be able to apply strategies learned in treatment to the home setting. Therefore, it is important to have family or caretaker involvement in the therapy plan. Ideally, this individual will also attend the treatment session(s). One of the most valuable intervention approaches to improve communication, when a person is facing more severe dysarthria, is for the listener to guide and control communication interactions.

Although cognitive changes will likely occur as the disease progresses it is important to not judge a person's intellectual abilities as more impaired because of reduced speech intelligibility. People who have HD are likely to comprehend much more than they can express.

General strategies that the listener can use to guide communication interactions

- Eliminate distraction.
- If you do not understand what was said, inform the speaker that you did not understand. Do not pretend that you understood when you did not.
- Pay attention to facial expressions or gestures to facilitate understanding.
- Ask for repetition or clarification, or repeat what you think was said in the form of a question. For example: “Did you say...?”
- You may need to go to an alternative approach by having them spell the word that was misunderstood.
- Allow the speaker enough time to convey his/her message.
- Be patient with the speaker.

General strategies that the speaker can use to improve verbal communication

- Speak one word at a time.
- Exaggerate each word.
- Speak at a slow rate.
- Start your voice at the top of your breath.
- Use short phrases, take a breath in-between phrases.
- Repeat the word or sentence when necessary.
- Rephrase if necessary.
- If having difficulty thinking of a word, try to describe the word.
- Indicate the first letter of a word that the listener does not understand.
- Use gestures, facial expressions.
- Use alternative communication techniques such as alphabet or word boards, picture boards, and/or electronic devices.

Severe Impairment in Speech/Language and Cognitive Function

Speech is considered to be severely impaired when it is no longer understandable. Unfortunately, in the advanced stage of HD, severe motor deficits limit the ability for the person to use other modes of communication such as writing, typing, and selecting words or phrases on a communication board. In addition, cognitive abilities may be impaired to the degree that the person is unable to learn new information. Because HD impacts cognition and motor control, it is often difficult or impossible for the person to use any type of complex augmentative communication system. Therefore, communication management relies heavily on a supportive communication partner who knows the person with HD well.

General strategies that the supportive communication partner can use to improve communication

- Use alphabet or picture board
- Provide choices
- Keep a routine
- Use large print calendars and memory aids

HDSA has a webinar on strategies that can be used to help with communication as cognition declines which can be found in the Living With HD section of www.hdsa.org.

Swallowing Problems Associated with Huntington's Disease

A swallowing disorder (dysphagia) is common among persons with Huntington's disease. Swallowing difficulties can result in food or liquid going into the trachea, often described as going down the wrong pipe, causing coughing, choking and, in the worst case, airway obstruction. Food and liquid entering the airway and settling in the lungs over time can result in aspiration pneumonia. Malnutrition and dehydration are also common in persons with untreated swallowing disorders.

Aspiration pneumonia is reported to be the most common cause of death among persons with Huntington's disease. Some of the more common swallowing problems associated with HD include:

- **Involuntary movements that disrupt the timing of airway protection ***

Airway protection occurs when your vocal folds close and your "voice box" or larynx moves up and forward so that food and liquid will be directed down the right path. If the voice box or vocal folds are moving unintentionally, aspiration may result.

- **Difficulty controlling the amount of food or liquid intake ***

When the person with HD takes too big of a bite or drink or adds more food/liquid before he or she has completely swallowed what was in the mouth.

- **Difficulty chewing or manipulating food ***

- **Delayed swallow reflex***

A swallow is made up of a voluntary stage (i.e. chewing and containing liquid or food in your mouth) and a reflexive stage. The reflexive swallow typically is when food or liquid contacts the “arch” or faucial pillars in the back of the throat. If the reflexive swallow is slow to start or delayed, the airway does not close in time, and food or liquid may go down the “wrong pipe” or trachea.

- **Holding food or liquid in the mouth and difficulty initiating a swallow***

- **Incomplete swallow where food or liquid remain in the mouth and/or throat***

- **Difficulty coordinating breathing with swallowing***

- Excessive belching

- Rapid, uncontrolled swallowing

Bolded problems that are marked with an * are particularly important to manage as they can increase the risk of aspiration, or food/liquid going into the lungs.

Eating is a pleasurable event for most people and social events often revolve around meals and traditional foods. When someone has a swallow disorder that prevents him or her from being able to eat or drink safely, it can be devastating for both the person with the swallow disorder as well as their family/caretakers.

How is Swallowing Assessed?

One of the major reasons for doing a swallowing assessment is to determine if a person is aspirating. Aspiration increases the risk of aspiration pneumonia. The amount of aspiration that will cause aspiration pneumonia is unknown. In fact, aspiration alone is not generally sufficient to cause pneumonia. However, the risk of aspiration pneumonia is significantly increased if other conditions are present including: being dependent on others for feeding, dependent for oral care, missing teeth, receiving tube feedings, having more than one medical diagnosis, taking multiple medications and smoking.

There are three types of swallowing assessments:

- 1. Clinical swallow assessment or bedside swallow assessment.**

During this portion of the swallow evaluation, the SLP observes and assesses the person with HD's swallow mechanism while he/she eats a variety of food and liquid consistencies. Based on information from the clinical swallow assessment the SLP may recommend swallowing strategies to reduce the risk of food and liquid going down the wrong pipe. In addition, the SLP may recommend that a more comprehensive swallow assessment be completed.

- 2. Modified Barium Swallow.**

This test, also called a cookie swallow, is done in Radiology. The person is asked to swallow different consistencies of barium, a chalky drink with the consistency (but not the flavor) of a milk shake. The barium is

viewed through an x-ray as it passes through the mouth, throat and into the stomach. This test takes 30-60 minutes. A barium swallow is generally a safe test, but like any procedure, there are occasionally complications which the doctor will discuss.

3. Fiberoptic Endoscopic Examination of Swallowing.

The SLP views the airway through an endoscope. A long, thin, tube with a lens on the end of the scope. It is connected to a light and a video camera. Images of the inside of the person's body can be seen on a screen. Endoscopy is minimally invasive and allows the doctor or Speech Pathologist to examine the inside of the throat. The whole endoscopy is recorded so that doctors and/or Speech-Language Pathologists can check it again.

When a person is found to be aspirating, it is difficult to know if they should continue to eat by mouth or if they should consider tube feeding for their nutritional needs. The goals of swallowing treatment and management are to maintain adequate nutrition and hydration through safe oral feeding for as long as possible, minimize the risk of aspiration, and improve or maintain the person's quality of life. Treating HD swallowing deficits can be especially difficult because the person with HD may not be a good judge of their swallowing problem, and the unpredictable choreic movements can affect the entire swallowing mechanism at any point in time. Therefore, it is important to have a swallow evaluation by an experienced speech-language pathologist that specializes in dysphagia to make recommendations on how to best manage the dysphagia.

Swallowing Strategies to Minimize the Risk of Aspiration

If the person with HD continues to eat by mouth, the SLP will make specific recommendations to minimize the risk of aspiration. Some common recommendations include:

Diet modification

Diet modification may include thickening liquids, pureeing foods, or avoiding certain foods. *HDSA's Family Guide Series, Nutrition and Huntington's Disease*, has more information about potential diet modifications.

Positioning

Positioning recommendations may include sitting upright at a table and keeping the chin down towards your chest as this position increases airway protection.

Compensatory strategies

Compensatory strategies include taking only small bites and sips, ensuring the mouth is clear of food before taking another bite, eating at a slow rate, supervision during meals to maintain a slow eating rate and appropriate bite size, and minimizing environmental distractions.

**** All caretakers should be trained in the Heimlich maneuver. You can learn about the Heimlich maneuver by doing a quick search online or by asking your primary care physician.**

Dysphagia Management Options

There are three different options when someone is experiencing dysphagia and is at high risk for dehydration, malnutrition, and/or aspiration.

Continue to eat by mouth

The person and/or his or her legal proxy may choose to continue to eat by mouth for quality of life reasons even when experiencing a severe swallow disorder. The person chooses to eat by mouth with a clear understanding of the consequences (malnutrition, aspiration, dehydration, choking, and possible death).

****To improve nutritional intake the person with HD should see a dietician. Often times the dietician will recommend taking supplemental drinks such as Boost™ or Ensure™. You can find a registered dietitian by going to eatright.org/programs/rdfinder/ and entering your code, or by clicking on the “Find a RD” button on the eatright.org home page.**

Tube feeding

Tube feeding can be done with or without oral feeding. The SLP and your physician can talk with you about the different options and the risks and benefits of each. Other professionals, including a social worker, chaplain, dietician, biomedical ethics committee, or nurse can also answer questions regarding the ramifications of placing a feeding tube.

There are two general types of tube feeding, a nasogastric tube and a G-tube. A

nasogastric tube is inserted through the nose and down the back of the throat and into the stomach and is typically used for very short periods of time. A G-tube requires a minor surgical procedure with insertion directly into the stomach and is generally used in longer-term situations.

No one right strategy works for everyone. It is important to discuss tube feeding well before it becomes necessary so that the person with HD can actively participate in the discussion.

Sample Cases

Case I

Demographics & symptoms

52 year old male with mid-stage HD has been living in a long-term care facility for 3 years. In the last month, staff noticed that he was coughing and clearing his throat when he drank clear liquids such as juice and coffee. He drank all of his liquids from a cup, threw his head back and coughed as the cup was emptied.

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 a clinical swallow evaluation.

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

Solutions

1. Person was given a straw. He sipped liquids with the straw and his coughing and wet vocal quality cleared. He now drinks all liquids, including coffee, via straw.
2. The straw keeps his head in a chin tuck position which is a position that is found to improve airway protection. It also helps him control the rate of intake.
3. He is given verbal reminders to take small sips. He is told that he can drink as much as he likes but needs to only take small sips.
4. The staff at the facility were educated about these strategies for safe swallowing and improved airway protection.

Case II

Demographics & symptoms

39 year old woman with early stage HD living at home. The caregiver reported that she was holding food in her mouth for long periods of time and that, even after she swallowed, there was still food left in her mouth.

Problems observed during a clinical swallow evaluation.

1. Chewing skills were ineffective.
2. Mouthfuls were too big.

Solutions

1. Person was placed on a finely chopped diet because the regular diet was too much for her to handle.
2. Cues were given to alternate sips of liquid with bites of food and to place less food on her utensil.

Case III

Demographics & symptoms

56 year old male living in a long-term care facility for 5 years. In the last several months, he has lost a significant amount of weight and is experiencing a lot of spillage of food and liquids when he eats.

Problems observed during a clinical swallow evaluation.

1. Severe dysarthria (muscles of mouth and throat weak).
2. Person holding head down when trying to swallow (leaning far forward).
3. Chewing skills were ineffective.
4. Person not coordinated for self-feeding anymore.

Solutions

1. Person was placed on a pureed diet and thickened liquids.
2. Positioned in a slightly reclined chair or bed for mealtimes.
3. Feeding now done by staff.
4. Family continued to bring soft favorites (snack cakes, candy) for quality of life.

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, healthcare professionals of different disciplines can help persons with HD to maintain independence and maximize quality of life. An early referral to a speech-language pathologist can help a person with HD and his or her family make a plan for the future. 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. You can learn more about team care for HD in the Living with HD section of hdsa.org. Email HDSA at hdsainfo@hdsa.org, or call call the HDSA Helpline at **888-HDSA-506** for information about resources in your area.

HELP FOR TODAY. HOPE FOR TOMORROW

HDSA Mission

To improve the lives of people with Huntington's disease and their families.



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