

**Huntington's New South Wales**



# **Communication and Huntington's Disease**

**Supported by NSW Health**



**Communication is the way in which we stay connected to our world. It puts us in touch with others, allows us to express ideas and feelings, give direction and exert control over our environment**

Listening, speaking, reading and writing are the traditional forms of communication. These skills of communication are greatly compromised, if not lost entirely in Huntington Disease (HD). All areas of communicative functioning are affected by HD. Because of the effect on communication and speech, the individual with Huntington Disease can be trapped in a world of their own, unable to communicate the most basic request or desire.

Although we are unable to stop the progression of Huntington Disease, we can manage the communication and speech difficulties associated with it to allow the individual with HD to maintain communicative skills for as long as possible. It must be remembered that the disease process can be as long as twenty years or more. Maintaining communication from the very beginning is essential. A speech therapist has a vital role to play in communication maintenance. Referrals can be direct, or go through the GP, the NSW Huntington Disease Service or the neurologist.

Speech therapists can play a major role in responding to the communication needs of the person with Huntington Disease at all stages of the disease process. The focus of the speech therapist with the person with HD is to evaluate and maximise their communication skills, augment those skills wherever possible and necessary, and guide and educate carers, family members and friends in the most effective ways to communicate with the person. The speech therapist will constantly reassess the changing communication needs of the person with HD, and plan effective management strategies throughout the course of the disease.

## **Speech and Language Difficulties Associated with Huntington Disease**

Communication, cognition (what we say and the thought processes needed to plan what we say) and speech production (how we say it) are all affected by Huntington Disease.

Speech production disturbances begin early on in the disease process. As the disease progresses, speech often becomes unintelligible and individuals are often non-verbal in the advanced stages of the disease. Speech difficulties include impaired breathing for speech production; hoarse, harsh, strained or strangled vocal quality; inappropriate rate, rhythm and pitch of speech (ie too fast, too slow, monotone, inappropriate stress on words); and imprecise articulation (ie unclear pronunciation of sounds).

Cognitive and language skills are also greatly affected by Huntington Disease. These deficits can include: difficulties beginning conversation; lack of spontaneity in communication; difficulty putting thoughts into words; reduced number of words available to the person; limited ability to respond within a conversation; specific word finding difficulties; difficulty understanding complex information; slow response time; impaired skills in reading and writing - ranging from physical difficulties to comprehension difficulties; problems learning new information and new skills; reduced short term memory skills; poor attention and concentration abilities; and lack of ability to organise, reason and problem solve.

All of these difficulties can be and usually are present in an individual with HD. They often begin in a mild form and become more severe as the disease progresses. Abilities are often unpredictable, because deficits occur randomly during the general progression of the disease. This unpredictability creates more coping difficulties for the

person with Huntington Disease, because they cannot rely on having or maintaining various skills at any given time. For example, the person with HD could make a request clearly at one moment, but then have tremendous difficulty articulating that same request a moment later.

Obviously the speech/language and cognitive difficulties associated with Huntington Disease are numerous and have a great effect on the individual's ability to communicate. In the later stages of the disease, many people are unable to communicate at all. However, even against what seem to be insurmountable odds, things can be done to help the person with Huntington Disease maintain the precious gift of communication for as long as possible.

In recent years, research has indicated that respiration irregularities and phonatory difficulties are the major factors in the problems people with Huntington Disease experience in speaking. (Respiration is the breathing support for speech production and phonation is the physiological process of air moving through the vocal tract and being transformed into sound within the larynx). This may be useful information for speech therapists developing treatment strategies, ie that therapy may be more beneficial if it is aimed at maintaining control over respiration and phonation rather than focusing on articulatory difficulties.

In addition to individual speech therapy, other carers can also use various techniques to facilitate and maintain communication for as long as possible. A number of suggestions follow.

## The Role of the Listener

One of the first guidelines in achieving effective communication with someone affected by Huntington Disease is for the listener to accept responsibility for the conversational exchange without controlling it. The person with HD can indeed use strategies to improve speech intelligibility and ability to participate in a conversation.

However, because of the many deficits (most particularly cognitive deficits), the person with HD must rely on external cues and guidance from the listener to achieve effective communication. Once the listener has agreed to accept this responsibility, communication has a much better chance of being successful.

Along with the listener being an active participant, the person with HD can learn strategies to improve communication and speech production skills. Much of the success of these strategies again depends on the listener's willingness and persistence in encouraging the patient to use these skills.

We must always be aware that even in the final disease stages the individual with Huntington Disease has something to share. Agitation and frustration and sometimes aggressive behaviour, can be the result of lack of communication. If the person with HD, like all of us, feels out of touch with others and out of control of their environment, a reaction of frustration will most certainly be the result. Aggressive behaviour is often the last avenue the person with HD has to exert control or to be noticed. Negative communication may be better than no communication at all.

## **Strategies for Communicating with the Person with Huntington Disease.**

The person with HD and the listener can use the following strategies to facilitate effective communication.

### **1. Ensure you have the person's attention**

If possible, establish eye contact before you start to speak. It is important that you have the person's full attention, so use "alerters" eg touch the person and say their name, or use a verbal lead-in such as *'I meant to ask you about .....*'. In this way, you don't begin with the important part of your message.

### **2. Use a reduced rate of speech**

Often when speech is difficult to understand, speaking more slowly helps. It is important for the listener to also use a slower than normal rate of speech, in order to model for the person with HD as well as to allow them extra time to process the information being presented. Emphasis of certain key or information carrying words or sounds may also help to increase understanding.

### **3. Repeat the message**

Repeating a message may be all that is needed to gain understanding. If the message is repeated several times, however, and is still not understood, it is best to try another strategy before frustration begins on the part of both the person with Huntington Disease and the listener.

### **4. Rephrase the message**

Saying something in a different way when it cannot be understood is another successful technique. This can involve

thinking of another word or phrase to communicate the same idea,

### **5. Simplify the message**

The main goal is to get the message across.

It is of little importance if speaker and listener use correct grammar or long involved vocabulary. The goal here is to simplify the message while preserving the communicative content. Use key words and clues such as pictures or real objects if necessary .

### **6. Spell part of the message**

Spelling a word or two in the message, may also facilitate understanding. This technique can be used verbally, in written form, or with a letter board on which the person points to the letters in the word. Again the key here is not spelling accuracy, but rather communication. To get the message across, the person may be more successful by spelling the word as it sounds, instead of using the correct spelling.

### **7. Identify key letters in the message**

Identifying the first letter of a word may be all that is needed to clarify the message. Here again, a letter board may be very helpful to identify the appropriate letter.



## **Strategies for the Listener**

### **1. Use yes / no questions**

Asking yes/no questions, and other types of very specific questions, which require short one or two word responses, may also help to clarify a message. This allows the person with HD to communicate their ideas without having to formulate and articulate long sentences. The listener, however, must accept responsibility in formulating specific and probing enough questions to understand the message.

### **2. Use cueing techniques to encourage word recall**

Word finding problems are often a common result of Huntington Disease. The listener can play an active role in 'cueing' the patient to help them recall the word. Cueing can include asking the patient to recall various attributes of the object, for example size, shape, location. Visually imagining (forming a mental picture) the object may also help to jog the memory. Specific questioning is also effective. These strategies will not only be helpful in word finding but also in facilitating memory skills in general.

### **3. Rehearse expected / common conversational exchanges**

General memory skills are also a problem. Practising commonly asked information, as well as maintaining a consistent routine and periodic orientation (eg reminders of activities/tasks the individual is involved in), helps to store things in long term memory. The person with Huntington Disease experiences the most difficulty with short term memory. Long term memory skills stay fairly intact.

#### 4. Combining information

Association of new information with old, previously-stored information also facilitates memory. 'Chunking' or combining the information will also help. For example, *'you have three things to remember about your trip to the doctor'*.

#### 5. Monitor perseverative behaviour and provide feedback

Perseveration (the continuous repetition of a topic or sentence and the inability to modify it after it has become inappropriate) is also a common problem among people with HD. Again the listener can help the person to monitor and change this behaviour. Some methods include:

- Reducing the number of times you change topics during a conversation, so that the person does not have to quickly go from topic to topic.
- Introducing a topic change by saying *'we are changing subjects now, we are talking about .....*'.
- Allowing some time to pass before introducing a new topic.
- Letting the person know when perseveration is occurring. The first step in changing behaviour is identification.

#### 6. Refocus person on topic

Along with perseveration, the person with Huntington Disease also experiences difficulty maintaining a topic of conversation. Again listener cueing, refocusing on the topic, and letting the individual know when they have gone off topic can help, as long as it is done sensitively to avoid further frustration. Minimising distractions can also reduce topic change.

## **7. Allow adequate time for communication**

Adequate time is also important for effective communication.

A person with Huntington Disease may require extra time to make themselves understood, and to formulate thoughts and ideas and express them. If you cannot spend the time, tell the person that you cannot. Something as simple as '*I don't have the time right now to talk but I'll get back to you as soon as I can*' will help.

As people with HD very often want what they want immediately, delaying the conversation until later should be avoided if possible, as aggressive behaviour seems to decrease when we respond promptly to requests.

However, when necessary, putting aside enough time (it may only be five minutes) will lead to much more effective communication than trying to talk while on the run.

## **8. Reduce distractions**

Reduction of environmental noise and distractions also plays an important role in effective communication with everyone, but most particularly with a person with Huntington Disease.

People with HD are often easily distracted by the environment. Reducing noise and distraction whenever possible gives the person the opportunity to focus and concentrate on the communicative task. The reduction of background noise will also help the listener to hear and understand the speech of the person with HD.

## **9. Recognise and use gestures and facial expressions as communicative tools**

Facial expression and gestures can also be very helpful tools, especially in the initial stages of the disease. Gestures and facial expression can be clues to help decipher what the person is trying to communicate. It is also helpful if the listener uses gestures and expression in order to provide the individual with Huntington Disease an extra communicative clue.

## **10. Try to gain topic knowledge**

It is very helpful to determine what the topic of conversation is early on in the discussion. This allows the listener to be able to ask more appropriate and specific questions as well as make more educated guesses when understanding becomes difficult.

## **11. Get the main idea**

In addition to topic knowledge, it is also important to remember that the goal is to get the main idea. Don't get bogged down in trying to understand every word.

## **12. Provide and ask for feedback**

While communicating, it is also important to periodically check with the person to make sure you have understood them correctly. Do not pretend to understand when you do not. This will only lead to frustration and mistrust. Repeating back the message, for example: *'Did you say ..... ?'* will help to clear up any misunderstandings.

## **13. Monitor listening and attending ability**

In order to effectively communicate, you must make sure the person is listening. Sometimes they may be hearing

you, even though it appears that they are not listening. Because listening and attending ability is decreased in Huntington Disease, you must check that the person is listening.

Communicating for short periods of time is often better than sitting down for a long stretch. It requires a lot of energy to effectively communicate, and that will be difficult to maintain for both speaker and listener for long periods of time.

If you as the listener feel that you are becoming distracted, stop and go over the previous information before continuing.

#### **14. Provide as predictable an environment as possible**

Reasoning and judgement skills are also impaired in Huntington Disease. Breaking down activities into small steps helps to compensate for this. Reducing demands and creating a predictable environment are also helpful coping strategies.

## Communication aids can be helpful

A word about communication aids such as letter boards, word boards and picture boards. Non-electronic aids tend to be the choice over electronic aids because the length of time the person with HD can use the board may be very limited.

Also, non-electronic aids can be made and altered at a moment's notice to meet the changing needs of the individual. At best however, communication aids tend to meet with limited success. They are often introduced too late in the disease process, when learning new skills is very difficult and reading and spelling skills are also severely affected. Word and letter boards are most helpful to those who can spell and/or indicate the first letter of a word. Picture boards have met with some success. The number of items on a board should be kept small to avoid difficulty with attention, distraction and scanning for the right picture. (See illustrations 1 and 2 for examples of communication boards).

**Please note, however, that boards must be designed individually to meet the specific needs of each person.**

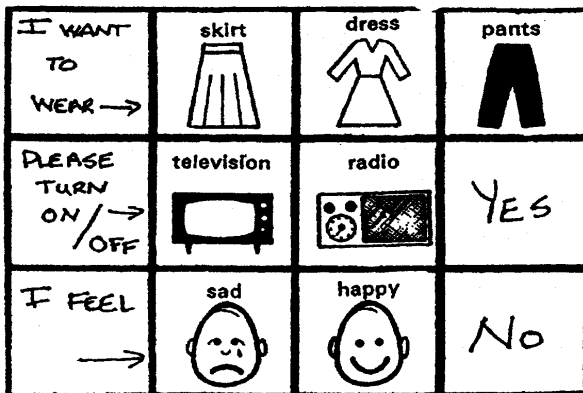


Illustration 1 Picture Board

I NEED THE NURSE	I NEED THE DOCTOR	I AM IN PAIN	I WOULD LIKE TO WATCH TV.	PLEASE CLOSE THE CURTAIN	I WOULD LIKE A DRINK.	I WOULD LIKE SOMETHING TO EAT	I WOULD LIKE TO REST NOW
I UNDERSTAND	I DONT UNDERSTAND	CLOSE THE DOOR	OPEN THE DOOR	PLEASE OPEN THE CURTAIN	I WOULD LIKE SOME TEA	I NEED THE WASHROOM	I WOULD LIKE SOME COMPANY
HELLO	HOW ARE YOU?	EXCUSE ME	MAYBE	YES	NO	OK	HOM
I THINK ITS TIME FOR A CHANGE	A	AN	HE / SHE	HIS / HER	THANK YOU	YOUR WELCOME	PLEASE
I	AM	HE	YOU	THEY	THEM	IS	ARE
DID	DO	DOES	BE	BEEN	I WANT	I NEED	GIVE

Illustration 2 Word Board:

For people in the earlier stages of the disease who have lost the ability to communicate verbally but still have cognitive/language skills.

Communication during later stages of the disease may very well be the only connection the person with HD has with their environment. They have lost control of their body and cannot take care of any of their own needs. Therefore maintaining communication at any level during this stage is extremely important.

Although the person with HD has suffered cognitive deficits and is non-verbal, he/she still understands a great deal of what is going on around them even though active participation is not possible. People affected by Huntington

Disease can remain aware and alert until the very end of the disease process.

Too often people with Huntington Disease are dismissed as not understanding anything because they cannot communicate verbally. With time and effort, you may well discover that you have underestimated how alert and aware the non-verbal person is.

### **Establish Communication Techniques with the Individual in the Advanced Stages of the Disease**

It is our responsibility as carers to maintain communication with the individual in the advanced stages.

1. A good approach here is to remember things that the individual previously asked for on a consistent basis and provide those comforts for them even though they are unable to ask for them.
2. Try to establish a routine of care with the person so that they know what to expect.
3. At all times explain what you are doing and why you are doing it. Use a soft gentle voice to avoid startling the person.
4. Talk to the person. Talk about the weather, the news, their interests, your interests - anything that will keep them connected to the world.
5. Do not treat the person as if they cannot share in communication. It may not be verbal. An eye blink, or a tiny smile may be your only response but it is well worth it. Maintaining communication allows the person at advanced



stages of the disease to go through the disease process with some dignity and provides them with every opportunity to communicate in any way possible.

If the person is able, you may be able to work out a series of movements to indicate basic needs. For example one eye blink for 'yes', closed eyes for 'no', raised eyebrows for 'thirsty' and so on. This may be difficult, however, if the patient is experiencing a great deal of uncontrollable movement.

6. Rely on family members to tell you more about the person so that you can have a better understanding of their history, personality and needs. Family pictures and other mementoes in the person's room will give carers helpful starting points for communication.

### **Let all involved know about the communication strategies being used**

It is important to let all who are involved in the care of the person with Huntington Disease know about the approaches that are being used to communicate. A good way to communicate with all carers is through bedside postings of techniques being used, difficulties the patient experiences, and any other pertinent but not confidential information.



## Fact Sheets

- Huntington's Disease in the Family – A Booklet for Young Children
- Huntington's Disease – A Fact Sheet for Teenagers
- Talking to Children about Huntington's Disease
- Presymptomatic Testing – The Consumer Experience
- Understanding Challenging Behaviour in Huntington's Disease
- Living with Someone who has Huntington Disease
- Making a Decision About Residential Care
- Guidelines for Meeting the Nutritional Needs of People with Huntington's Disease
- Eating and Swallowing Difficulties in Huntington Disease
- The Importance of Dental Care in Huntington Disease
- Communication and Huntington Disease

*These Fact Sheets are available from  
Huntington's NSW  
Please see the back page for contact details.*

## Helpful Contacts

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For further information on Huntington Disease, a membership form, or details of HD services in NSW, please contact Huntington's NSW

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Huntington's Victoria