

Huntington's New South Wales



**Eating and Swallowing Difficulties
in Huntington Disease**

Supported by NSW Health

Eating and Swallowing Difficulties

Difficulties with eating and swallowing (dysphagia) and maintaining a constant body weight are among the most troublesome complications of Huntington Disease (HD).

There are many factors involved and many reasons why these problems occur. These include:

Changes in Appetite

Most people with HD have voracious appetites. They always seem to be hungry and have a tendency to cram food into their mouths to try to satisfy their hunger, causing problems with choking and loss of food by spillage. Behavioural problems can often be attributed to the severe hunger of the person, which has not been duly met by the diet. This problem is a common occurrence when speech and communication problems are present and the carer finds it difficult to understand what the person is trying to say and therefore may not be aware of his/her need to eat. Appetite can be affected by a number of other factors such as the fear of spilling things, fatigue, changes in the diet, and irritation over the difficulties experienced when eating and drinking, all of which can make eating a frustrating, upsetting and even confrontational time. Although hungry, the person with HD may therefore actually refuse to eat very much food at all.

Choreiform Movements - Problems of Chewing and Facial Control While Food is in the Mouth

Feeding and eating difficulties arise from the choreiform movements of the face and neck, incomplete lip closure and irregular movements of the diaphragm. The loss of fine muscle control and co-ordination can make eating a tiring and frustrating experience.

Independent eating is also made difficult by the involuntary movements of the upper body and difficulties with fine motor control and co-ordination, making feeding oneself increasingly difficult.

Deterioration of the Muscles Involved in Swallowing

"Swallowing problems are rarely a complaint at the time of diagnosis of Huntington Disease, however, as the disease progresses, the co-ordination of the swallowing mechanism becomes more and more impaired. Along with poor co-ordination, protection of the airway is also compromised due to sudden unpredictable gulps of air during the inhalation cycle of breathing. When inhalation occurs, the vocal cords are open and the airway is exposed, creating a high risk for aspiration of food particles into the open airway".

(Estelle R. Klasner, "Managing Swallowing Difficulties Associated with Huntington's Disease", Huntington Society of Canada)

Choking frequently occurs during eating and drinking, and is attributed to the abnormalities present in several phases of ingestion:

The anticipatory phase: this includes the decision before the food enters the mouth. The person with HD can make an inappropriate food selection and is incapable of monitoring rapidity and quantity of food intake. Cognitive impairment rather than chorea is responsible here.

Mastication or chewing: choking occurs here because the person with HD often 'squirrels' food in the mouth until there is too much food to swallow safely.

The actual swallowing: sometimes there is a too rapid or immediate attempt to swallow which triggers coughing and choking.

Weight Loss

More calories are needed. Weight loss in HD has been long observed and frequently attributed to the involuntary movements. The cause of the weight loss is still unclear and many theories have been put forward. One theory is that the weight loss may be a manifestation of an underlying biochemical defect.

Whatever the cause the fact remains clear that a greater calorie intake is required.

Because this is a problem relating to each individual, we recommend that a dietician be contacted who will work out an appropriate diet plan. Ask your own GP to refer you to the dietician in your area or seek advice from the Dietician at the NSW Huntington Disease

Service (see back page for contact details). Even with an increase in calorie intake, the problem of weight loss may not be solved. Many people with HD do not maintain their body weight and become emaciated, suffering the consequences of tissue breakdown and increased susceptibility to other illnesses.

Changes in Mood

As previously mentioned, the presence of anxiety or depression can affect the person's appetite. Anxiety and stress often trigger visible chorea and can therefore affect the swallowing process. The actual fear of choking, drooling or the embarrassment of making a mess while eating can actually increase the chance that it will happen. Many people with HD show great resistance to alterations to their life-style, which can make it very difficult to introduce the correct food into the diet.

Symptoms That May Indicate Swallowing Difficulties

An individual with Huntington Disease may experience only a few of these symptoms:

- Collection of food at the side or on the roof of the mouth
- Spitting food out of the mouth
- Long delay between food being placed in the mouth and swallowing
- Excessive tongue movement
- Difficulty coping with different food textures or liquids
- Coughing and choking
- A 'gurgly' voice quality
- Excessive secretions from the nose or dribbling
- Regurgitation of material through mouth or nose
- Deterioration of speech and voice
- Weight loss
- Recurrent episodes of pneumonia

(Extracted from 'Swallowing Difficulties and Speech', Margaret Pozzebon in "Caring for Persons with Huntington's Disease", 2nd edition, 1990)

The Selection, Preparation and Arrangement of Food

The selection, preparation and arrangement of food are important in most cultures. Many of our memories of family life and friends are tied to eating, food and drink. A pleasant family meal is an important part of that family's life, a chance to talk, show love and concern, play roles in the family, catch up on the news. Yet this important part of family life can become a frustrating, tearful, angry time when someone is unable or unwilling to select, prepare and eat food in the usual way. Someone who for years has shopped, cooked and enjoyed food may now feel lost and worthless because all or some of these things are difficult, tiring or even impossible.

Anything that can be done to make food selection, preparation and eating retain its place is worth consideration. The benefits are too important to lose.

The following information includes general principles and practical skills that should be carried out to make the whole feeding process less frustrating for the individual, family and other carers.

A comprehensive assessment and treatment plan devised by a speech pathologist and occupational therapist is important to facilitate independence and safety for the person with HD. Speak with your GP or staff at the NSW Huntington Disease Service for assistance with this.

It should be remembered that nutrition is an important consideration in the health care of any person in subsequently preventing the development of a secondary illness.

1. Preparation for Eating

Before meal times, be prepared to prevent distractions happening during the meal.

Assemble the appropriate feeding aids within easy reach of the dining area. Prepare food that is easily swallowed and of the correct consistency and texture. The consistency of a stiff mousse or porridge is often easiest to swallow.

Position the chair that the person with HD will sit on whilst eating, away from the wall, out of the corner and near the outside of the

table so that the person is readily accessible should choking occur. To help prevent fatigue occurring during mealtimes it is advisable that the person with HD rests for half an hour to two hours before eating.

Passive limb exercises performed prior to meals can reduce tension caused by anxiety, which can often help to minimise the choreic movements.

2. Mealtime Aids

There are many aids available that, although fairly simple in context, can make a mealtime the pleasant time it was always meant to be.

The Independent Living Centre has a substantial collection of aids to assist people with disabilities with all the activities of daily living. An occupational therapist is also able to provide advice about eating aids. The occupational therapist, outreach nurse or dietician at the NSW Huntington Disease Service are also able to help you.

As there are many aids available to meet some of the needs of people affected by Huntington Disease, it would be impossible to mention them all in this booklet. However, the following information gives a summary of useful aids and some hints on how to use equipment that is already available in the average household:

- Plastic cups, plates and bowls are light in weight, durable and easy to manage when there is poor control of movement.
- Scoop bowls and plate guards can be used to stop food being pushed off the plate.
- A preheated hot plate maintains the warmth of the main course throughout the meal, an important necessity as the person with HD requires more time to eat and this ensures that the meal remains warm and appetising.
- Cutlery is easier to hold if the handles are large and "fat". You can build up handles yourself, using rubber and taping this in place.
- Some brands of bicycle handlebar grips fit ordinary household cutlery, the "fatter grip" can lessen choreic movements. Some people have improvised their cutlery quite satisfactorily using rubber hair rollers or the foam type.

- A hand strap or grip can be added to cutlery. The strap, often of Velcro, fits around the fingers and makes the utensil easier to grip and hold during eating
- A double-handed cup, preferably with a spout, is an important piece of equipment to aid drinking.
- Fill any cup or mug only halfway to cut down on the chance of spills.
- Flexible straws or straws with one-way valves assist drinking.
- Larger straws are available for thicker substances.
- A non-slip mat will stop the plate, bowl or cup moving during eating. A damp face-washer placed under the plate has proved equally useful.
- Plastic tablecloths can be attractive, useful, and easy to keep clean. Also, warm plates will not slip on a plastic cloth.
- Plastic aprons can also be useful. There are many available that are not expensive. Look out for those with pockets so that small items can be carried safely.

3. Positions for Eating and Drinking

Never administer fluids, solids or medications while the person with HD is lying down or in a reclining position.

Maintain a secure, comfortable and upright position throughout the meal. The person should be seated upright in a chair at a 90 degree angle, and then asked to lean slightly forward. The feet should be flat on the floor and the elbows can be put firmly onto a waist-high table using the arms and palms of the hand to steady the body. This position helps to reduce and stabilise the choreic movements.

Throughout the meal the head should be kept slightly down and forward. Throwing the head back whilst eating and drinking increases the risk of gagging or choking. If the head is unsteady, steady it by placing the palm of one hand to the person's forehead or behind the neck or back of the head.

If the person with HD has to remain in bed for mealtimes, use a sitting

position with the head of the bed elevated to a 45-90 degree angle, using pillows or a foam wedge placed under the mattress, and place at least three pillows horizontally behind the shoulder and neck.

The carer should be seated in front of the person they are feeding in his/her clear field of vision.

4. Environment During Meals

Even the smoothest run household can be chaotic at mealtimes. Sometimes such a situation will make the person with HD irritable, anxious and unwilling to eat. Perhaps a quiet meal before or after the rest of the family, with companionship available, will be the only answer. However, the following information about the environment during meals is worth keeping in mind when assisting with someone's diet.

Ensure the surroundings are as attractive and pleasant as possible. A little soft music from a radio or a favourite record can help the mood of the occasion, dining therefore being made as much a social as a nutritional event.

Avoid emotional situations which interrupt the swallowing process and cause difficulties controlling secretions.

Coughing and choking are exacerbated by tension.

Maintain an unhurried environment free from too many comings and goings.

5. Schedule For Meals and Breaks

The feeding process should be at least half an hour to an hour in duration. Eating can be a tiring experience so allow as much time as necessary for mealtimes. Even if you have a thousand other things on your mind or things to do, make sure you look as if you "have all the time it takes" and that you do not regard the assistance as a chore to be done as quickly as possible.

It is always a good idea to plan for smaller and more frequent meals, particularly when meals seem to be lasting too long; for example, instead of having three large meals a day, try four to six smaller ones supplemented by high calorie drinks. Schedule high calorie snacks and nutritional breaks between main meals. Serve smaller portions with refills so that the task of eating seems less daunting. Postpone eating if the person with HD is tired or upset. Restart at a more convenient moment.

6. The Diet of the Person with Huntington Disease

Generally speaking, the diet of the person with HD should contain foods that are easy to chew and digest, are nourishing, varied, easy to swallow without difficulty and provide for adequate vitamin, mineral and calorie intake.

When the person is newly diagnosed he/she does not usually have too much of a problem eating and a normal everyday balanced diet with a slightly increased quantity is adequate. This is a good time to ask your GP to arrange a consultation with a dietician.

Normal weight can be recorded and arrangements made for further periodical advice. As feeding difficulties present themselves, pay particular attention to the type of foods eaten. Foods particularly easy to swallow are soups, sauces and soft scrambled eggs. It is a good idea to add cream, butter or full cream milk. For variety use ice cream and yoghurt. The thick and creamy yoghurt is the right consistency.

For further suggestions contact the NSW Huntington's Disease Association (see back page for details) for a free copy of "*Guidelines for Meeting the Nutritional Needs of People with Huntington Disease*", by Karen Keast.

Foods to avoid :

- Food that is transformed into a poor consistency by incomplete oral movement such as white bread, or bacon which becomes stringy. Tomato and other skins are often difficult to remove from the hard palate with the tongue which lacks co-ordination.
- Small, coarse and hard foods that may be inhaled accidentally such as peanuts, potato crisps, hard toast. Bread and butter put in the oven for a few minutes makes a good substitute for toast.
- Avoid over-spiced and sharp food.
- Take poultry and meat off the bone.
- Avoid fish with bones unless filleted completely.
- Avoid shredded vegetables like coleslaw, carrots and lettuce.
- Avoid mixing food textures. Never give the person a mouthful of fluid to wash down the food. The two differing textures are confusing in the mouth and invariably the person gags and chokes.

Drinks often create the greatest problems. If drinking water causes choking, try using chilled water, or if this doesn't help, avoid giving

water. Drinks that are too thin in texture do present problems. Try adding an enriched cereal or rice cereal to thicken soups, drinks and sauces. Juice that is already thick, like tomato and pineapple juice are easier to swallow. Liquids with 'bits' in it, like minestrone soup, are particularly difficult to manage.

Coffee, tea and soft drinks given on their own without some modification should really be avoided. They are just too thin and non-nourishing to be worth it.

If, however, they are real favourites, use them as an ingredient in a shake or thicker drink, or accompany them with a muffin or roll for "dunking".

Carob added to drinks and soups acts as a thickener and does not alter the flavour of the dish.

It has been found that tea made with tea leaves causes more problems than if made with tea bags. The small tea leaves that escape the strainer stick to the hard palate and can cause choking.

7. Eating the Meal and Tips on Feeding Someone

As the disease progresses, it is probable that the person with HD will become unable to self-feed. This loss of a basic self-care skill can be very hurtful to the person and can result in all sorts of reactions to the need to be fed or to the need to eat at all. If you are sensitive to these feelings, much can be done to make the task easier and more pleasant for both the carer and the person eating.

Before starting the meal, and in between courses, offer the person a few sips of iced water, which can often assist swallowing.

Both the carer and the person with HD should be comfortable and relaxed. Face the person you are feeding.

Throughout the meal, the person should be encouraged to eat slowly and chew the food well.

To make swallowing easier:

1. Encourage the person to breathe out before placing food into the mouth;
2. Using a small spoon or fork, take a small amount of food and place it into the person's mouth;
3. Ask the person to close his/her lips tightly together as soon as the cup or spoon has been taken away;
4. Ask the person to breathe in and out through his/her nose; ask the person to chew the food, then pause;
5. Ask the person to stop breathing and swallow. You cannot breathe and swallow at the same time.
 - Stroking the larynx whilst the person is swallowing encourages laryngeal elevation and therefore actually helps swallowing. Gently massaging the face and neck muscles at the upper jaw angle just below the ear before and during a meal can be helpful towards relaxing the person with HD and assisting the swallowing process.
 - If the person feels that the food is sticking in his/her throat, ensure that the food is of the correct consistency and the texture is not too dry.
 - Encourage the person to cough and try to clear his/her throat between courses and at the end of a meal.
 - Do not introduce more food into the person's mouth until the previous mouthful has been swallowed.
 - Allow for frequent rests and pauses throughout the meal. Always watch for signs of fatigue or anxiety and recommence feeding at a more appropriate time.
 - Encourage assistance from the person who is eating. Perhaps they could manage drinking from a straw by themselves or holding small pieces of finger food.

8. Activities After Meals

After meals the person with HD should sit up for fifteen to thirty minutes to aid digestion and avoid aspiration.

Perform or encourage oral hygiene (cleaning of teeth etc.) after all meals and snacks to prevent aspiration of food particles that might be inside the person's mouth.

Be able to evaluate the amount of food consumed at the end of each session, to ensure adequate nutritional intake.

9. Taking Medications

Tablets when in the form of capsules or enteric coated (sugar coated) are the easiest to swallow.

If pills are difficult to swallow crush them and place in custard or some other dessert which can be easily swallowed.

Most medications are more easily administered in a thick liquid form. Talk with your pharmacist and seek his/her advice about such preparations.

10. Coping with Constipation

There are many causes of constipation so always check with your GP first. Some of the main causes are:-

Lack of exercise

The normal function of the gastrointestinal tract is impaired by immobility and hence constipation is *often* a problem.

Lack of fluid

A normal diet should have at least two to three litres or 6 to 8 glasses of fluid a day.

Lack of fibre in the diet

Increasing the daily intake of fruit, vegetables, wholemeal cereals, prunes and prune juice will help. If this is not satisfactory, add natural bran to foods such as puddings, stews and soups. It is possible however to add too much fibre to the diet so check with your GP first. Hot lemon water stimulates the bowel and can often relieve constipation.

Constipation has been found to be the source of some faecal incontinence and diarrhea particularly when the person has had a long history of constipation. Have a word with your GP if such a situation should arise.

NOTES

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

Huntington's New South Wales

PO Box 178, West Ryde, NSW 1685
Tel: (02) 9874 9777
STD Free Call: 1800 244 735 (Country NSW)
Fax: (02) 9874 9177
Website: www.huntingtonsnsw.org.au
Email: info@huntingtonsnsw.org.au

NSW Huntington Disease Service

Alison Anderson (02) 9845 9957
Speech Pathologist, Westmead Hospital

Karen Keast (02) 9845 9956
Dietician, Westmead Hospital

Jet Aserios (02) 9845 6699
Social Worker, Westmead Hospital

Cecelia Lincoln (02) 9845 6699
Social Worker, Westmead Hospital

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

**We recommend that Carers obtain a
First Aid Qualification so as to be
able to respond to a choking incident**

Acknowledgements: Huntington's Disease Association
(England & Wales)
Huntington's Victoria