Managing Swallowing Difficulties Associated with Huntington's Disease

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Foreword

Over the past two years, April 1987 to April 1989, I have worked half-time as a speech pathologist at Runnymede Hospital in Toronto and half-time in a research capacity, funded by the Huntington Society of Canada.

During this initial research phase, I have undertaken three preliminary investigations. They are:

1. Acoustic characteristics of the Dysarthria (speech problems) associated with Huntington's Disease;
2. The Efficacy (Benefits) of Speech Treatment with Huntington's Disease;
3. Swallowing Disorders and Huntington's Disease.

The Huntington Society has asked me to continue my research into speech and swallowing difficulties, and I will begin this new phase, one and one-half days per week, in early 1990.

In the meantime, the Society has asked me to prepare two booklets, one on Swallowing and one on Communication, which outline practical stategies for management of problems in these areas.

Because of time and funding constraints, we were not in a position to compile separate booklets for family and for professional caregivers at this time. Family members may therefore find more technical information than they require in the introductions, and professionals may find the format slightly different from what they are used to.

I also realize that many families and care facilities may not have the opportunity to request the comprehensive swallowing investigations I have proposed. In these situations, I suggest that a speech/language pathologist be consulted and a clinical evaluation be performed. This can be done at home, in an outpatient clinic or any other facility. Speech/language pathologists can be contacted through local health care facilities, public health or home care programs, or the Huntington Society may be able to recommend speech/language services in your area.

I am excited by the initiative and foresight of the Huntington Society of Canada in addressing the serious difficulties associated with communication and swallowing problems in HD. I am also encouraged by the Society's commitment to further investigate management strategies in these areas.

I welcome additional ideas from both family and professional caregivers. Your comments may be directed to me through the Huntington Society's National Office, Box 333, Cambridge, ON N1R 5T8.

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For the convenience of those who have purchased **A Manual For Care**,
this booklet has been printed in three-hole punched format, so that it can be inserted in *Chapter IX Food and Eating*.

First published in 1990 by the Huntington Society of Canada.

Introduction

Huntington's Disease (HD) is a genetic neurological movement disorder. HD has been identified as a principal neurological cause of swallowing disorders (Groher, 1984). The extensive neuropathological involvement associated with Huntington's Disease causes all of the anatomical structures involved in the swallowing function to be affected by the disease.

The act of swallowing food and fluids is a complicated mechanical process. It requires precision and accurate timing of many muscle movements. When these abilities are compromised, the ability to swallow food and fluids safely and effectively is also challenged. Many physical and psychological concerns often result from swallowing difficulties. The Huntington's Disease individual and his/her family are naturally very concerned with the basic function of swallowing. Fear of choking and the possibility of not being able to eat and/or drink can be overpowering. Frustration can occur when the individual experiences difficulty eating and this may lead to a refusal to eat at all.

The Normal Swallow

In order to fully understand the impact of Huntington's Disease on the act of swallowing, a basic understanding of the physiology of the normal swallow is extremely helpful. As mentioned earlier, the act of swallowing is a complicated neuromuscular event requiring accurate coordination and timing of muscular movements.

Swallowing is a combination of voluntary and reflexive movements, which ordinarily takes no more than three seconds (Dereiko and Stout, 1986). The process of swallowing can be divided into **four phases**: 1. oral preparatory phase, 2. oral phase, 3. pharyngeal phase and 4. esophageal phase. (See Illustration 1) The **oral preparatory phase** begins when food is placed in the mouth and is chewed. The food is mixed with saliva, and the consistency is altered in order to prepare the food for swallowing. The teeth, tongue, lips, jaw and cheeks all assist in this preparation process. Phase two, the **oral phase** of the swallow, involves the gathering of food by the tongue and palate and moving the food to the back of the throat. The tongue holds the food against the hard palate until the swallow reflex begins. Both the oral preparatory phase and the oral phase of the swallow are voluntary in nature. The third phase, or **pharyngeal phase**, begins as food enters the pharynx. During this phase, the pharynx contracts in order to help food travel down the pharynx, and the vocal cords close to protect food from entering the airway. The esophageal sphincter (a ring of muscles) also relaxes in preparation for food to enter the esophagus. The final phase, or **esophageal phase** of the swallow, begins as food passes from the pharynx into the esophagus. Muscle contractions take place to allow the food to travel through the esophagus and into the stomach. Both the pharyngeal phase and the esophageal phase of the swallow are considered to be automatic (involuntary) in nature.

Exact timing and precise coordination are extremely important for swallowing to occur in a safe and efficient manner. Difficulty in any of the phases of swallowing can severely compromise both the efficiency and safety of the swallowing mechanism.

The **involuntary movements** and **decreased muscular control** caused by Huntington's Disease affect both the coordination and timing of the swallow. HD affects every phase of the swallowing process. Several other factors associated with Huntington's Disease also affect the safety and efficiency of the swallow. These include **appetite changes**, **memory changes**, and general **changes in cognitive functioning** (the way individuals process information and think).

Swallowing and Huntington's Disease

Swallowing problems are rarely a complaint at the time of diagnosis of the disease. But as the disease progresses, swallowing function becomes greatly impaired and not only poses a daily threat to life, but is often the cause of death due to repeated occurrences of aspiration pneumonia, a type of pneumonia that is caused by ingesting foreign objects (such as food particles) into the lungs. Aspiration pneumonia can occur when the swallowing function is impaired, because the airway is not adequately protected and food particles can enter into the lungs during swallowing. In addition to swallowing difficulties, Huntington's Disease patients often develop an increased appetite which places a further focus on food intake. Cognitive changes often impede the individual's ability to learn compensatory swallowing strategies.

Difficulty swallowing, increased appetite requirements, and decreased cognitive abilities pose a challenge when providing swallowing management to the Huntington's Disease patient.

As previously mentioned, swallowing difficulties rarely present as a clinical sign at the time of diagnosis of Huntington's Disease (Groher, 1984). However, recent videofluoroscopic investigations indicate that subtle changes are present in swallow function before clinical indications become apparent (Klasner, 1988 in progress).

Initially, individuals with Huntington's Disease may begin to lose some of the skills required for independent eating. The ability to handle a fork and knife becomes increasingly impaired. Food choice is an effective management strategy at this stage. Individuals will often choose finger foods, such as sandwiches and other items that do not require a great deal of manipulation, in order to maintain independence in eating ability.

As the disease progresses, and involuntary movements become more apparent, the ability to swallow and the coordination of the swallowing mechanism become more and more impaired. (See Illustration 2.) Along with poor coordination, protection of the airway is also compromised due to sudden unpredictable gulps of air during the inhalation cycle of breathing (Groher, 1984). 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. Increased difficulty with tongue coordination and movement impairs the ability to form a bolus (food that has been gathered by the tongue and formed into a "ball" ready to be swallowed) and position food material correctly in order to initiate the swallow reflex. Food and fluids can often enter the pharynx before protective mechanisms have been put into place again, creating a high risk of aspiration. Food material is also often pushed out of the mouth by involuntary tongue movements. Because of poor tongue manipulation, the entire timing of the swallowing sequence is compromised: either initiation of the swallow occurs too quickly and/or the swallow reflex can be significantly delayed as the tongue attempts to gather food material and form a bolus in order to trigger the swallowing reflex.

It appears that the most impaired stages of swallowing for the Huntington's Disease individual are the first two, i.e. the preparatory stages of swallowing, where food is being prepared to be swallowed. During these stages, the individual experiences the most difficulty, and this affects subsequent stages of swallowing. Clinical signs which may indicate that an individual is experiencing swallowing difficulties include: coughing and choking at mealtimes; chronic pulmonary congestion; poor chewing ability; regurgitation of food; refusal of certain foods; spitting of food; inability to swallow voluntarily; and a lack of, or weakened, cough reflex. Huntington's individuals, as well as all their caregivers, must be alert for these indications at all times.

Although the swallowing difficulties of the Huntington's Disease individual are complex and all-encompassing, management strategies can be implemented at many levels to maximize efficiency and safety of the swallow.

First and foremost, caregivers involved with the Huntington's Disease individual must be aware of the great potential for swallowing difficulties, choking and aspiration that are associated with Huntington's Disease. They must be alert at all times to the clinical indications of swallowing problems, be prepared to recognize these indications and seek out professional services, and be prepared to provide appropriate food material to minimize any risk. Caregivers should also be trained in emergency techniques, should an incident occur. (See *Appendix B* for emergency techniques.)

Illustration 1
Normal Swallow

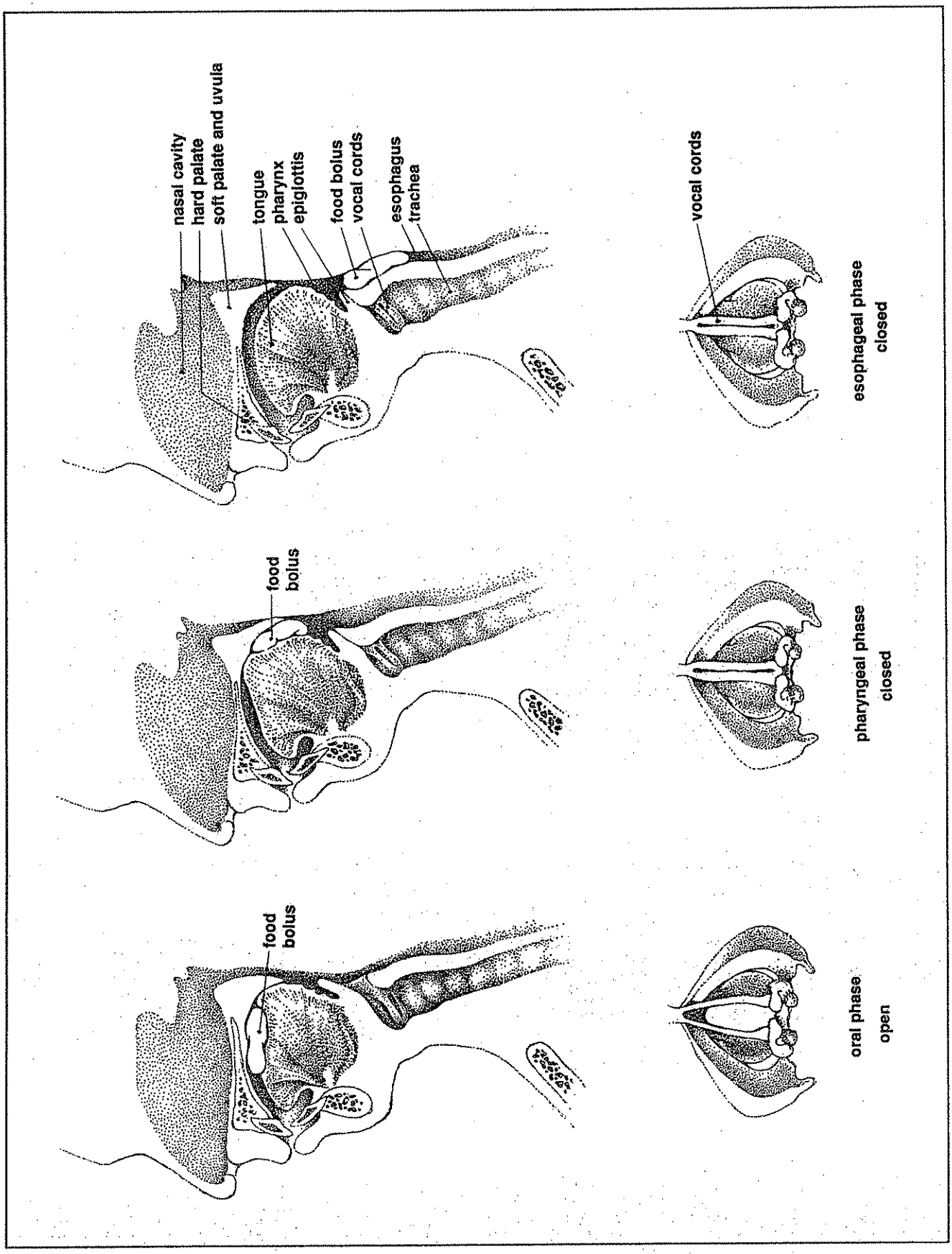
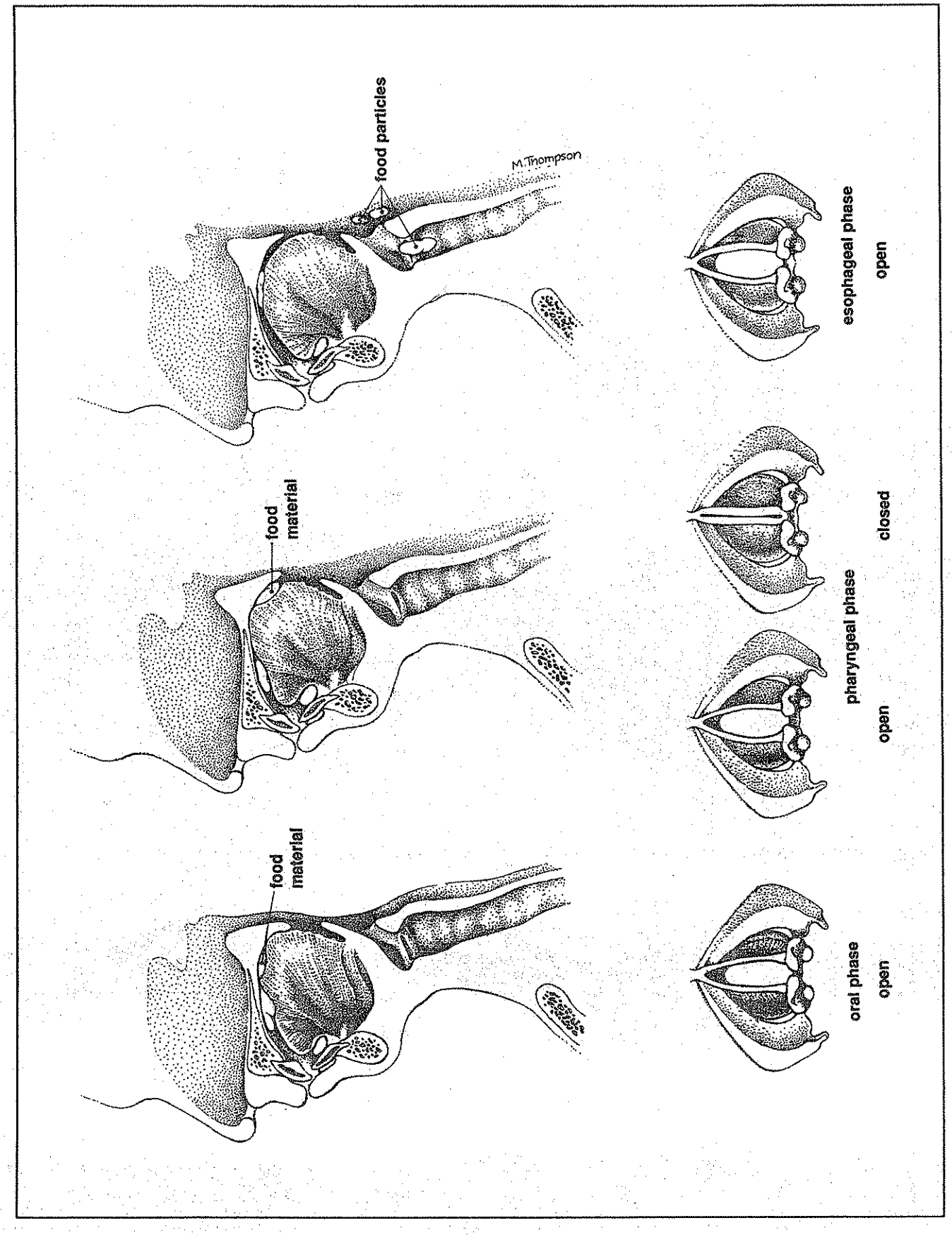


Illustration 2

Possible Difficulties During Impaired Swallowing



Management

Step One: A Professional Assessment

In order to provide effective and successful management of swallowing difficulties with the Huntington's Disease individual, it is critical to have a speech pathologist assess the individual's swallowing situation. The local public health nurse or home care program coordinator can help to locate a speech pathologist. If the person resides in a care facility, it is essential to obtain a comprehensive interdisciplinary assessment of the swallowing disorder. Because the act of swallowing involves many processes, it is important to obtain an extensive evaluation which looks at the swallowing disorder from many varied viewpoints. It is not enough to only consider physical difficulties; one must also consider nutritional needs, cognitive status, psychological well-being and many other issues, which can be effectively addressed from an interdisciplinary standpoint. A suggested interdisciplinary team may consist of the following members:

The Individual: It is extremely important to understand the individual's needs from his/her point of view. We must also be aware of the individual's level of awareness of his/her swallowing problem, and make him/her an active participant in his/her care for as long as possible. Even if the person with HD does not provide much input, it is important that he/she be included in decisions about his/her care.

Primary Caregiver: If the individual is still at home, it is essential to get an idea of the caregiver's awareness of the swallowing difficulties as well as his/her ability to follow through with the program prescribed by the speech pathologist and/or team. If the individual is in a care facility, the family caregiver may still have an important role to play in supporting the management plan and should be kept fully aware.

Nursing: If the individual is living in a long term care facility, it is often nursing that first notices swallowing difficulties and makes the referral to the swallowing care team. Nursing can provide an accurate history of the individual's eating habits and major difficulties. The nurse will often be the primary individual who will carry out the swallowing management plan that has been recommended.

Speech/Language Pathologist: A speech/language pathologist often serves as the coordinator of the swallowing care team, and directly assesses swallowing, communication and cognitive abilities of the patient. She/he is also responsible for designing and implementing a treatment program involving various facilitation techniques (positioning, food consistency, direct therapeutic techniques, etc.) to manage swallowing difficulties. She/he trains nursing staff and other direct caregivers in feeding and facilitation techniques, and constantly re-evaluates and changes the program according to individual needs. She/he may order diagnostic tests, such as videofluoroscopy (a videotaped x-ray of the swallowing process in motion) to gain objective information.

Occupational/Physio Therapist: Evaluates positioning ability and fine motor abilities of the individual, in order to provide information regarding positioning during mealtime and the ability to maintain independent feeding skills. Can also help to evaluate the use of specialized feeding equipment.

Dietician: Provides appropriate meals in terms of consistency and evaluates nutritional needs of the patient on an ongoing basis. Monitors food and fluid intake.

Social Worker: Provides background history of the individual and family, and insight into the individual and family's needs and attitudes. Also provides ongoing support.

Physician: Makes decision concerning oral/non-oral feeding, overall medication management, and provides ongoing reinforcement of swallowing treatment plan.

Dentist: Is often consulted concerning dentition of patient, poor fitting dentures and overall dental care.

Any other health care professional can become a team member involved in the individual's overall care.

Step Two: Management Strategies

After the health care team has evaluated the situation, individual management strategies can then be implemented. Management strategies should be constantly reviewed and re-evaluated to meet the changing needs of the patient. Because HD is a progressive disorder, re-evaluation is particularly important.

Following a complete swallowing assessment, the health care team can develop a swallowing care strategy to facilitate safe and effective swallowing. The following suggestions may facilitate swallowing abilities:

1. Awareness by all Caregivers of Swallowing Difficulties

All caregivers involved with individuals experiencing swallowing difficulties must be aware of the difficulties and the swallowing care plan at all times to ensure safe and effective swallowing. Changes should not be made in the swallowing care plan without consultation with the speech pathologist and/or health care team.

2. Minimize distractions

Due to cognitive changes, individuals with Huntington's Disease can and do become easily distracted. Attention is needed in order to concentrate on the act of swallowing. Concentration on the swallowing process allows the Huntington's Disease individual to exert some control over the swallowing mechanism and thereby play an active role in ensuring a safe and effective swallow.

3. Encourage a slow, relaxed eating style

Often, because of increased appetite, individuals with Huntington's Disease tend to eat in a very rapid manner, which can create a higher risk for choking and aspiration. It is important to encourage a slow eating style. A good way to do this is to involve social interaction during mealtime and offer one or two food items at a time. Mealtime is often a social event and this interaction should be encouraged without interrupting concentration during swallowing. The use of a teaspoon rather than tablespooon also encourages a slower eating pace.

4. Positioning of Individual during and after mealtime

The position of the patient during and after mealtime can have a great effect on the individual's ability to swallow and can also serve as a method of preventing aspiration. Individuals should be in an **upright** position with head tilted slightly downward during mealtime, and should remain upright for 30-45 minutes following meals to prevent regurgitation and aspiration of food. Modified seating may have to be utilized to ensure appropriate positioning during mealtimes.

5. Carefully evaluate and monitor food consistencies

Food consistencies (i.e. how thick or thin food/fluids are) can make a tremendous difference in an individual's ability to swallow. Because Huntington's Disease usually involves discoordination of many muscular movements needed for swallowing, particularly tongue and jaw control, it is important to provide food consistencies which the individual can successfully swallow. Pureed items are not always the best choice, because they may be too thin for the tongue to gather. Thicker items of a pudding-like consistency are often effective, because these items are easier to gather in the mouth. The consistency of many foods can be easily altered. For example, soup can be thickened by potato flakes, pablum, or the addition of corn starch. The Huntington's Disease individual may be able to eat many regular food items with minor modifications and should not be immediately placed on a pureed diet once swallowing problems become apparent. A blender or food processor is invaluable in creating the food consistencies necessary for the individual to eat safely. It is necessary to re-evaluate the individual's swallowing function on an ongoing basis so that food consistencies can be adjusted accordingly.

6. The drinking of thin liquids must be carefully considered

Thin liquids such as water, coffee and juice are by far the **most difficult** items for the tongue to gather and hold in a bolus before the swallow reflex is triggered. Because of poor tongue coordination and a possible delay and/or early initiation of the swallow reflex, the consumption of thin liquids often poses a major threat to the swallowing-impaired individual. Poor tongue coordination and impaired timing of the swallow reflex can lead to fluids entering the pharynx and the airway before protective mechanisms have been put in place. **Thickening agents** such as THICK-IT (available from Oetker Foods*) have been used successfully to thicken thin liquids to a nectar, and/or honey-like consistency. This facilitates swallowing without removing the pleasure of drinking. If THICK-IT is unavailable, substances such as gelatin or jello may be used to thicken fruit juices. Contact a dietician at your local hospital or public health department for more information on other available thickening agents. Before using any thickening agents, an evaluation must be done to assess need and consistency requirements.

* Oetker Food Company, 2229 Drew Road, Mississauga, Ont. L5S 1E5, Attention: Peter Blair. The Ontario toll-free number is 1-800-387-6939. Outside Ontario, call (416) 678-1311.

7. A dry mouth can cause swallowing difficulties

Many medications that may be prescribed by the physician for the Huntington's individual can cause a dry mouth. Saliva is a natural lubricant which is used to facilitate the swallowing process. If the mouth is dry, swallowing will become very difficult. Making sure the mouth is moist before beginning a meal can ease the act of swallowing considerably. Starting the meal with a liquid may moisten the mouth sufficiently.

8. Placement of food items must also be considered

If the Huntington's Disease individual is no longer able to feed him/herself and is being fed by a caregiver, the caregiver must be aware of appropriate placement of food items in the mouth in order to stimulate efficient swallowing. Functioning of the oral cavity needs to be assessed so that food placement can be appropriate. The placement of food will depend greatly on tongue strength and coordination. Generally, food can be placed in the middle of the tongue and a slight downward pressure with the spoon will aid in triggering the swallow reflex.

9. Coughing should be encouraged

Coughing is the natural protective mechanism when **choking** occurs. If the individual is experiencing choking, encourage vigorous coughing in order to alleviate the difficulty. Often, a good cough is all that is needed to clear the airway. Of course emergency procedures should be implemented when necessary. (See *Appendix B* for emergency procedures.)

10. Mealtimes should be limited to 25-30 minutes

Timing during a meal is important. If a meal takes more than 30 minutes to complete, it is likely that the individual and caregiver are experiencing fatigue and possible frustration, thereby creating an atmosphere for aspiration to occur. Meals should be completed in 30 minutes. Allow the individual time to rest and, if necessary, reintroduce food at a later time, when both individual and caregiver are rested and alert.

11. Mouth should be clear of residue after eating

The mouth should be thoroughly checked for remaining food residue after mealtime, either by the individual or the caregiver. If residue remains in the oral cavity, there is a possibility it could be aspirated long after mealtime is completed. A clean mouth after mealtime decreases the chance of an occurrence of aspiration.

12. Specialized feeding equipment

In order to maintain independence during feeding, specialized feeding equipment should be considered. Rimmed plates, plate guards, non-slip mats, scooper bowls, specialized forks, spoons and cups can ease

gathering of food. Many items are available on the market. (See *Appendix A* for resources. You may also refer to section 1X g 70-72 of the Huntington Society's **Manual for Care** in regard to mealtime aids.)

An occupational therapy assessment will be most helpful in choosing appropriate equipment. Occupational therapy assessments can be obtained from any hospital that has an occupational therapy department. If the hospital does not have an occupational therapy department, they may be able to refer you to an occupational therapy service. These assessments may also be available through the public health nursing office or home care program.

13. Maintain independent eating skills for as long as possible

With appropriate food choices (eg. finger foods, fruit, sandwiches, etc.), appropriate food consistency, and the use of specialized feeding equipment, independent feeding skills should be maintained for as long as possible. This is of utmost importance to the Huntington's Disease individual, particularly when he/she may have had to relinquish other activities of daily living to caregivers. Maintaining independence in eating allows the Huntington's Disease individual to retain some control over his/her environment and partake of many social events involving food for a longer period of time (going to parties, restaurants, etc.). Eventually the need to maintain adequate nutrition will outweigh the need for independent eating — often a very difficult decision to make.

14. Supplements can be an effective way to maintain adequate nutrition

In the past few years, several commercial nutritional supplements have become available. Although expensive, it may be helpful to use these products to ensure adequate nutrition. Supplements can serve an excellent purpose by providing nutrition without a great deal of preparation. Supplements can be thickened easily to any consistency, and can serve as a quick way to provide additional nutrition. A caution however — supplements should be used only as a supplement to a well balanced diet and should not be used as the only source of nutrition. See *Appendix A* for suggested nutritional supplements.

15. Therapy to retrain swallowing function

Various therapeutic techniques are available to retrain the swallowing function. In order to benefit from this training, the individual must be cognitively aware at all times and have intact judgement and problem-solving abilities. This may not be an alternative for the Huntington's Disease individual, given that cognitive deficits are part of the disease process and it is often difficult for individuals to learn new things. If cognitive skills are intact at the time of the assessment and retraining the swallow is chosen as a therapeutic option, cognitive skills must constantly be re-evaluated for the possibility of deterioration as the disease progresses. It must also be remembered that swallowing abilities will also change as the disease progresses, therefore therapeutic techniques also have to be monitored and constantly changed to address the changing needs of the individual. Swallowing retraining techniques should be performed by a qualified speech/language pathologist.

16. Syringe/Bolus feeding should not be considered

Syringe or bolus feeding is **not** an appropriate feeding technique. This technique involves placing pureed food items in a syringe and injecting the syringe into the individual's mouth. This method of feeding does not allow the individual to prepare for the act of swallowing, and therefore protective mechanisms may not be in place and aspiration of food can readily occur. This method of feeding also does not allow the individual to take an active role during mealtime and should not be considered as an option.

17. If adequate nutrition cannot be obtained orally, alternative feeding methods will be considered

If the individual cannot obtain adequate nutrition orally, it may be necessary to consider alternative feeding methods, such as the gastrostomy tube. This is a decision that must be thoroughly discussed with the physician, other members of the health care team and the person with HD.

Conclusion

Swallowing difficulties associated with Huntington's Disease are complex and challenging. However, creative, practical treatment strategies **can** be implemented to maximize safe, efficient swallowing and allow for the Huntington's individual to enjoy mealtime for as long as possible. A **comprehensive assessment and treatment plan** devised by a speech pathologist or an interdisciplinary health care team is of utmost importance in order to provide the Huntington's Disease individual with the best management strategies possible. Although it is not possible to stop the progression of Huntington's Disease, symptomatic treatment can provide management strategies which allow the individual to enjoy as active and full a life as possible, for as long as possible.

Bibliography

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Elliot, J.L. "Swallowing Disorders in the Elderly: A guide to diagnosis and treatment." Geriatrics, Vol. 43, January, 1988.

Groher, M.E. Dysphagia Diagnosis and Management. Butterworths, Boston, 1984

Logemann, J.L. Evaluation and Treatment of Swallowing Disorders. College-Hill Press, San Diego, California, 1983.

Appendix A

Specialized feeding equipment can be obtained from:

1. Convalescent supply stores in your area

2. Maddak Inc

3. Fred Sammons Inc.

Catalogues can be obtained at convalescent stores for both of these companies.

Nutritional Supplements:

Recommended supplements include:

Enrich with Fibre
Ensure
Ensure Plus
Ensure Puddings
- available from Ross Laboratories

Sustacal Puddings - available from Mead-Johnson

Resource
Resource Plus
- available from Sandoz Nutrition

These supplements can be obtained at larger drug stores — if not in stock, drug stores will order products for you.

Appendix B

CHOKING

You may want to cut out this section and put it someplace that is handy when you most need it. Review the procedures until you feel you could carry them out quickly and effectively if something happens; don't wait until the need arises. Practice on someone if that helps to make the routine familiar to you, and make sure that family members all know what to do and feel prepared to do it. Instruct the person with the disease, since they can take measures when alone to help themselves, if they stay calm.

You may feel safer if you copy the basic instructions onto a little card for your pocket when visiting, or for having at hand in various parts of your home. If you eat in restaurants, you will be reassured to know that the Canadian Restaurant Association has been carrying out a wide program to train restaurant staff on what to do when someone chokes. Most staff know what to do, and can do it quickly to help.

A: IF HE/SHE CAN BREATHE A LITTLE, TALK

Stay calm, reassure.

- encourage big, deep coughs rather than shallow, irregular ones

B: IF HE/SHE CANNOT BREATHE, IS SILENT

Stay calm, reassure. Begin the Heimlich manoeuver (see illustration next page).

- bend the person over, head near the knees; stand a little to one side and support the person's side with your own body and arm with the heel of your other hand, or the edge of your hand held firm, give four sharp, short blows between the person's shoulder blades
- repeat
- if the person is still not OK, stand behind him and put your arms around the body at the level of the pit of the stomach, just at the bottom of the ribs
- put your two hands together as one fist, and draw up sharply and hard in a sort of "bear hug" to dislodge the bit of food; repeat if necessary

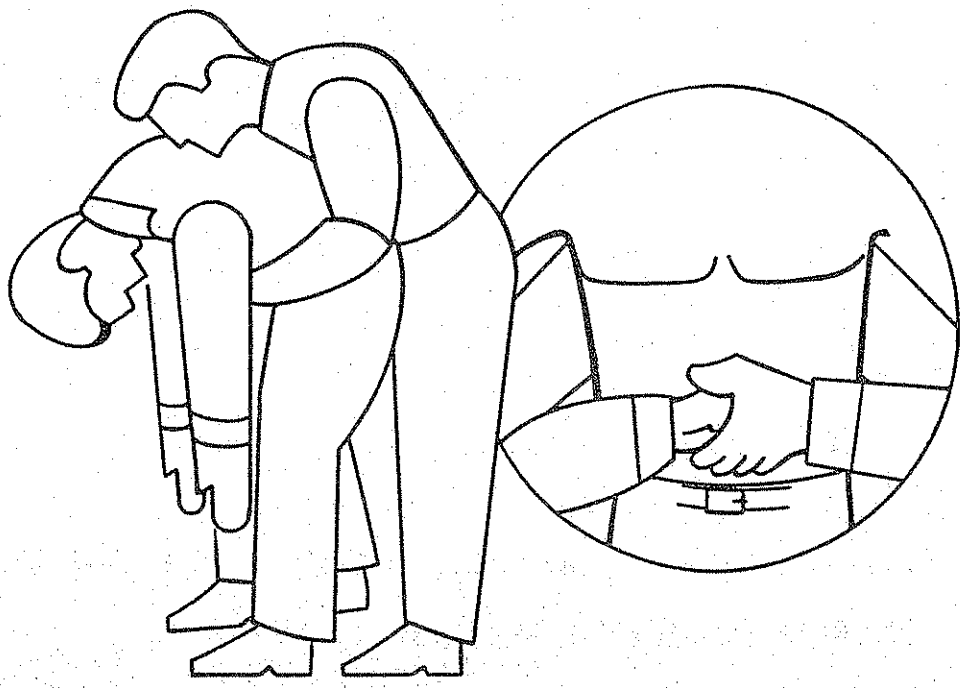
WHEN THE EPISODE IS OVER, TRY TO REASSURE IN A CALM VOICE. The experience of choking is a very frightening one.

TRY TO FIGURE OUT WHAT CAUSED THE EPISODE SO THAT SUCH AN EPISODE CAN BE PREVENTED IN THE FUTURE.

CHOKING

HEIMLICH MANEUVER

Rescuer standing, victim standing or sitting



OR Rescuer kneeling, victim lying face up