

Nursing Care In Late Stage Huntington Disease

By Joanne Hunt, RN

Interest and Involvement of the Author

I graduated with a diploma in nursing from Victoria General Hospital School of Nursing in 1993. I am presently working on my Bachelor of Science in Nursing through distance education from St. Francis Xavier University. I worked for four and a half years with Northwood Homecare caring for many clients with different illnesses acute and chronic, mostly palliative care. For the past 4 years I have worked at the Nova Scotia Hospital in acute care, Short Stay and Assessment Services working with clients acutely ill or in crisis situations and completing comprehensive admission assessments.

As a health care provider, person at risk for HD and a care giver of a family member with HD, I have become more involved with trying to educate family members and co-workers about HD. I feel I should begin by sharing why I am so interested in teaching health care workers about Huntington Disease. I experienced in my own work environment how people respond to persons with HD when they do not understand the disease itself.

I have done a lot of teaching with them as my colleges to help them understand about the process of HD and help them feel more comfortable caring for some of our clients.

Thus helping the client receive the proper and needed care and understanding at a stressful time in their life. I feel I have a lot to share with others due to my own personal experience in caring for my grandmother who has HD and is presently in the late stages.

I want people to understand and care about what persons with HD and their families go through and feel comfortable with them. As with my family history I may be dealing with my mother developing HD some day and even possibly I may be the person with HD on the receiving end of care some day too.

So I think by teaching what I know will help others with HD, their families and the caregivers caring for them.

I Hope You Will Be Able To Use This Information In Caring For A Person With HD. Remember. We Are All Human Beings.

Nursing Approaches to Care In HD

To start I would like to begin with the basic principles, we as healthcare givers use in all of our practices. These are especially important to remember when working with persons with Huntington's Disease as not all the same practices will work with everyone.

Persons with Huntington's Disease are normal, healthy, high functioning individuals until they begin showing symptoms of the disease and then they slowly begin becoming more and more dependant on others for their daily activities and eventually loose total control of their lives, needing others to do for them.

The basic principles to guide any of our interventions we as health caregivers undertake are as follows :

1. The client is entitled to the best quality of care.
2. Everyone is entitled to a sense of hope for a better life.
3. The client is the center of our care.
4. We as caregivers must be flexible, innovative and always keep an open mind.
5. Value your own style in what you do, but also value that of others. Your personality is your most valuable asset, so use it.

Some philosophical hypothesis based on contact with persons with Huntington's Disease and their families to maintain optimum care modalities are as follows :

1) Persons with Huntington's are capable of living a satisfying life during the course of the illness.

- Caring takes precedence over curing when as in Huntington's there is no cure available. So the care provided to a person is important to aid the individual and their families to live a close to normal and enjoyable life as possible. Caring is necessarily total, comprehensive, holistic and balanced.

2) Persons with Huntington's Disease are capable of using retained abilities, and learn new living skills and strategies.

- Their abilities they have need to be maintained and optimized and are more relevant then working with the irreversible disabilities. Within a structured routine they are able to learn to do tasks as a repetition, day in and day out and in this way they can learn to do a task in a more modified way that they are capable of at the stage of illness.

3) Improved quality of life will have a beneficial effect on the course of Huntington's Disease.

- The quality of life is more to be preserved than the quantity of life, the better quality of life the more enjoyable a person with Huntington's and their families can have together and a more fulfilled life.

4) Comprehensive care for Huntington's Disease persons will have , in the long term, benefits for families.

- As I have already discussed, caring is necessarily total, comprehensive, holistic and balanced. Huntington's is a disease not only of an individual, but of a whole family as it has an impact on a family's dynamics. A family's dynamics changes but when care is taken with aid and assisting the whole family, everyone will benefit.

5) The community and the health profession can be educated, using Huntington's Disease as a model, to place more emphasis on caring medicine.

6) Comprehensive care in Huntington's Disease can be used as a model for developing innovative health care delivery to the neurologically disabled and the elderly.

Loss of control, power and autonomy is a major issue in Huntington's disease, as this is a neurological disease in which there are cognitive, emotional and physical changes. But one important thing to remember is that a person with Huntington's does know what is going on around them.

Remember they were a normal, healthy, high functioning individual that slowly loses control of their movements, actions, ability to attend to their personal care and eventually needing to depend on a person or persons for all their activities of daily living.

As an individual they are noticing themselves how they are losing their abilities and this is a scary and emotional period for them. We as health caregivers have to in different ways depending on the stage of illness help the person with Huntington's Disease feel they still have some control of what is going on in their lives. We always have to give them some power and decisions to make.

One important way to do this is to always maintain a structured routine for them, ensuring to implement it for them making changes only when necessary and allowing the person a choice, remembering to give a choice of one thing or another. Not more than two things to choose from.

As the disease progresses more changes would need to be made to the routine, or at least how things in the routine are performed. This is what takes a lot of time, patients and working together with the client and family to see what works best for everyone. So we as caregivers need to help the client feel more in control therefore more powerful.

What is Power ?

Knowledge and information bring power

So it important to get to know your client and family and what has been problems and issues for them.

Respect is power

Always respect your clients and their families, and put yourself in their position, this helps you to respect them more. This is their lives and how they have to live so respecting them and their wishes will give you respect in return.

Communication is power

Always tell your client what the plan is, next in the routine, explain what you are going to do before you do it and while you are doing it. Also remember to always listen to what your client is saying, watching especially for their nonverbal cues as well. They need to know that what they think and say matters and that you are listening to them.

Making choices is power

Allowing a person to choose between one item or another, such as wearing a blue shirt or a red shirt, is a way of empowering them. They get to decide which of the two to wear, which helps them feel in control, they decided and nobody told them what to do.

And ...

Receiving help to maintain control is power

Before getting into specific care needs for persons with Huntington's Disease, I thought I would share with you a sample of nursing diagnosis in late stage Huntington's Disease.

- **Alteration in regard for safety.** Especially of self safety, this being compounded by impulsiveness, unrealistic expectations and problems with memory.
- **Alteration in mobility.** Related to uncoordinated movements.
- **Alteration in nutrition.** Related to swallowing and choking which causes fears around eating and drinking.

- **Alteration in swallowing.** Related to impaired voluntary control of mouth, tongue, swallowing mechanisms, respiratory control and impaired judgment.
- **Alteration in self care.** Related to uncoordinated movements and cognitive changes.
- **Altered and impaired judgment** and poor impulse control related to cognitive changes.
- **Alteration in emotional comfort.** Which may be a result of behavioral manifestations, for example depression, loneliness, anger, fear, explosive outbursts and alienation.
- **Alteration in communication.** Especially speech which results in difficulty being understood and in turn results in powerlessness, frustration and alienation.
- **Alteration in social relationships.** This is a family disease in which there are enormous changes in the "normal " family life.
- **Alteration in elimination.** Incontinence is an eventuality in the late stages.

Personal and Physical Care Needs

In Huntington's Disease there are cognitive changes and impairments. The cognitive areas that affect a person's performance include their speed, reasoning, planning, judgment, decision making, emotional engagement, preservation, impulse control, temper control, perception, awareness, attention, language, learning, memory and timing. Overall this affects how a person with Huntington's Disease attends to their activities of daily living and how they respond to their personal environment.

Caring for a person with Huntington's can be very challenging at times due to the cognitive changes in the brain. They may request things to be done now and if you do not attend to their need immediately they may become agitated, disruptive and more demanding.

Sometimes in the long run, it can be more efficient for you to do what they want right away, as to prevent a more disruptive, agitating behavior to progress. This type of behavior may also occur when the client has a problem with preservation or getting stuck on a thought or request, and asking you to do the same thing over and over and over again. Such as positioning their pillow as it is never right for very long or even asking for a cigarette.

These types of behaviors become more and more common in advanced stages of Huntington's Disease. Both can be managed in a similar way, by setting a routine and certain rules that all caregivers and family stick by. If you are unable to attend to them right away give them an exact time that you can meet with them such as in fifteen minutes or at six o'clock, and most importantly follow through with your set time.

Do not tell them " in just a minute " or " When I'm finished " as a minute to them is just exactly sixty seconds, and they will continue to reapproach you, which in turn frustrates you and the client. Attending to these simple requests in a routine, calm, supportive manner helps the client to feel good and even make them happy.

Do not pass off their disruptive, demanding and agitated behavior as bad behavior as this is part of the disease, this is due to cognitive changes in the brain.

Other behaviors which may result due to lack of impulse control and anger over their lost independence maybe hurtful comments and profanity directed towards family and caregivers. This type of behavior can be very hurtful even though as family and caregivers we understand it is part of the disease.

You always have to remember not to take this type of behavior to heart, continue to be polite and supportive. Many times you will even find that the person with Huntington's Disease will apologize for their behavior outbursts.

By understanding the cognitive changes in the brain, you as caregivers can begin to find causes of inappropriate behaviors. It may initially seem like a difficult complex situation that is out of control, but in the end there is a very simple solution, set routines and stick to them. And always remember to listen to what your client is saying, whether it be verbally or in their non-verbal cues.

Huntington's Disease is also a movement disorder with a presence of involuntary movements which most persons with Huntington's are not bothered by and may not be even be aware of. These movements are worsened by anxiety, stress and depression, while decreased during sleep and vary with posture and positioning.

There is also an impairment of voluntary movements which correlates more with functional disability as they have slow and uncoordinated fine movements and some rigidity.

So basically, persons with Huntington's Disease have difficulty and eventually are unable to plan, laying out or gathering items needed to complete a task such as brushing their teeth and unable to complete the steps of the usual everyday task. The cognitive issue effects planning and organizing the task while the movement disorder effects physically completing the task.

It is typical for a person to loose interest in attending to their hygiene so establishing a self - care routine is important and give no more assistance in care than is needed. Encourage tasks by prompting or giving cues.

Safety and Acceptable Risk

An incident relating to mobility and acceptable risk I would like to share with you has to do with a Huntington's client I had in my practice, for an admission assessment.

My report I had received prior to her being transferred to our facility was that she was acting out, aggressive and difficult to manage, so they medicated her with 2 mg of Lorazepam. So when this client arrived by ambulance she initially was verbally aggressive to me.

I approached her in a calm, pleasant manner, encouraging her to relax as I just wanted to ask her a few questions, her primary caregiver was also present with her. She got off the stretcher with the help of her caregiver and sat in a chair demanding a coffee. I made her a coffee as she liked it and put it in a plastic handled mug we had on the unit, as the styrofoam cup was too unsafe for her, due to her quick and constant movements. I gave her the mug and she voiced pleased with me taking the time to consider a mug being easier for her.

She requested to go to the bathroom and did not want to go in the wheelchair but instead to walk herself. Of course there was the increased risk of falls due to her movements and recently being medicated, but there was no reasoning with her. She walked to the bathroom with her caregiver on one side and I followed behind with the wheelchair. She did do well at that time walking on her own. After finished in the bathroom she walked back to the waiting room in the same manner.

I completed my assessment with her and then it was time for her to meet with myself and the doctor. She again wanted to walk to the interview room on her own. As she would be more influenced by the lorazepam by this time I encouraged her to allow me to assist her getting up and walking. Again she refused, wanting to do it herself.

Her caregiver and the doctor went across to the interview room while I stayed with my client for standby assistance. She got up easily, but as she started to take her first step she started leaning backwards. I acted quickly and grabbed onto her shirt and pants, as she was falling away from me. But I was able to keep her from falling and hitting her head on the wall.

She looked at me and then asked me to help her and she took my arm. She voiced to me that this scared her and was thankful that I allowed her to do it herself but more thankful that I did not just walk away but was there to catch her.

I allowed my client to do as she wished as to aid and encourage her independence, even though I knew she was at a very high risk to fall. By allowing her to try I empowered her. I used the acceptable risk and she appreciated this and thanked me, especially when I helped her from falling.

As the disease has been progressing there are many safety issues that had to be put into place, assessments of safety issues and also safety precautions have continually needed to be put into place.

The safety issues all become so important with the progression of the disease as the movement disorder worsens, the cognitive abilities continually changing, thus putting your client at a high risk for harm.

One issue with a person with Huntington's Disease that causes concern is their delayed response to pain, hot and cold sensitivity, these being important issues when dealing with everyday activities such as moving about even in bed, eating, bathing and for many, smoking.

There are a lot of persons with Huntington's Disease that refer to smoking as " one of the last pleasures I have left ", thus smoking becomes symbolic of independence for them. Nicotine also does have a noticeable effect in helping decrease some of their movements.

This can cause problems between the smoker and their caregivers as a person with Huntington's Disease poses a safety risk. This being related to behavior changes such as disinhibition, personality changes, impaired judgment and diminished capacity for self observation, and simultaneously the impairment in voluntary movements increase in the chorea movements.

As people with Huntington's Disease often have a diminished sense of hot and cold and often burn their fingers when lighting their cigarettes or smoking it down to the butt. Due to their impaired judgment they become unaware of the danger of burning themselves or the floor.

So basically to aid in allowing a person their pleasure, a plan needs to be put into place. Your plan can include setting certain times in the day for smoking only in designated areas. Please do not allow them to smoke in bed. A good setting would be in a room with no rugs on the floor, wearing flame resistant clothing and supervised.

Other things you may try earlier on in the disease progression , is discussing with your client and their families about nicotine patches or the use of Zyban to aid in smoking cessation.

The main importance for you as a caregiver is to help keep your client safe in all aspects, but this does not mean limiting their pleasures, it is important to be accommodating and flexible to help them stay safe from harming themselves.

Mobility and Seating

As Huntington's Disease is also a movement disorder there becomes changes in mobility that begins that begins in early and middle stages of the illness and continues into late stages.

Some of the noticeable physical changes in mobility is the wide - based dance - like gait, giving uneven weight bearing. This type of gait is related to hyper - extension at the hips which results in a more swaying gait and gives high risk chance to falling backwards, though there can also be flexation at the hips which rapidly causes accelerated walking in which the client is unable to control and gives increased risk of falling forwards.

Persons with Huntington's have a decrease in arm swing, have difficulty initiating walking and moving sideways and backwards is often difficult. Surprisingly, many people whose gait is so severely effected that they appear to be a danger to themselves and others have an amazing capacity to recover their balance when necessary, and some may function better if they walk alone, rather than with assistance. This does not mean that they do not need assistance or someone supervising their walking as they may still actually fall.

Some have difficulty judging distances and in differences in appearances and texture of floor surfaces, which may make them hostile going through doorways or moving between furniture.

This is where acceptable risk needs to be taken into consideration in order to help the person with Huntington's maintain their independence. It is important for caregivers, the client, their families and the doctor discuss the risks of allowing the person with Huntington's Disease to continue walking.

Even though there is such a high risk for them falling and getting hurt, not allowing them to mobilize will actually hurt them more by taking away their feelings of independence, thus taking away a part of their. It also increases their lose of muscle tone.

Persons with Huntington's Disease already know the progression of the disease will eventually take away their freedom of mobility but we as caregivers do not need to take it away too soon. A person with Huntington's Disease hangs onto their ability to walk as once they lose it, they know their disease is progressing to a more later stage.

As long as a person with Huntington's can walk unassisted or assisted we should allow them to do so. This needs to be discussed with everyone involved especially when a caregiver fears that if their client falls and injures themselves they will be in a situation of a lawsuit. Acceptable risk is allowing the client to do something that you know is a high risk activity.

As the disease progresses the movements worsen and the person with Huntington's Disease will not only have more and more difficulty ambulating but also more difficulty sitting in a

regular chair, even to the point of with the movements throwing themselves out of a chair or falling out.

A regular wheelchair or gerichair in late stages are not quite appropriate for persons with Huntington's as they need chairs with padding and angled seating such as a Broda semi reclining chair, which can be wheeled about and is fully padded to prevent injury. See pictures of the Broada on the last page.

The angled and padded chairs such as a Broda chair and Q- foam chairs are especially adopted for persons with Huntington's Disease and often reduce the need for other restraints, as using restraints on a person with Huntington's can cause more injury due to their increased chorea movements. This does not mean that no types of restraints can be used, seat belts are usually all that would keep a person in their chair.

Ensure all restraints are padded and used properly and as minimally as possible. Do not try to restrict all their movements as with the movement disorder when a person with Huntington's becomes stressed or anxious the movements worsen and if restraints are restricting any chorea movements your client can actually sustain an injury.

So minimize restraints as much as possible, they should be used only for safety not for difficult behavior or trying to control difficult behavior.

The angling of the Broda and Q-foam chairs is also a very helpful tool for the positioning of your client, as when positioned properly and comfortably, a person with Huntington's can be more relaxed, thus have a decrease in movements.

The padding and angling of the chair also aids in decreasing the chance of pressure sores, especially on the hips, buttocks and coccyx, as in these chairs the pressure is taken off these areas.

With the progression of Huntington's Disease and movements which eventually decrease mobility to a point where a person with Huntington's is at too high of a risk to walk anymore. This is the point in the disease when the person with Huntington's becomes more and more dependant on others for total care.

They will need assistance with positioning while in bed, assistance up to the bathroom, commode or to use the bed pan, assistance with transfer from their bed to a chair, increased assistance with personal care and eating.

This does not mean that they just go into bed and stay there, it is important to still get a person with Huntington's up in a comfortable chair and repositioning, just because they are unable to do most things for themselves does not mean we give up on them and leave them in bed all the time.

They will need help more than ever with doing their range of motion exercises and positioning to prevent and contractures. Your client will need more assistance overall and it is important to have someone help them that they can depend on and feel confident in.

Some Tips To Prevent Falls

- Do not call out to a client from behind, which would cause them to turn around abruptly and possibly lose their balance
- Do not interrupt a person abruptly
- Do not give medications while they are standing and do not try to feed them standing up
- Do not step in to try to prevent client from "bouncing off the wall" this could actually cause them to fall
- Proper footwear is very important, in the later stages high top sneakers or light weight workboots are more likely to be helpful
- Most common places for falls are in front of a chair, a toilet and/or a bed
- Try "touch-turn-sit" strategy
- Orient client to their surroundings and teach about any peculiarities of any of the equipment in their surroundings
- Encourage your client not to get up or do transfers without someone present

Environmental Changes to Prevent Falls

- Stabilize the furniture so it can not move
- Use chairs with armrests and high backs
- Clear away any unused furniture, providing clear accessible paths
- Remove any throw rugs and thick carpeting
- Rearrange tables and lamps away from most frequently used paths
- Place padding on frequent bumped furniture and doorways and/or have them rounded off
- It is estimated that a person with Huntington's Disease needs twice the space than that of an average individual, thus it is important for no clutter and free of hazards
- Side rails on a hospital bed
- Tub rails and tub seats
- Raised toilet seat

Swallowing and Nutrition

I have already discussed that as Huntington's Disease progresses the involuntary movements become more severe and this affects the ability of swallowing and the coordination of the swallowing mechanism. There is a higher risk of aspiration of food particles as inhalation may occur when the vocal cords are open and the airway is exposed. This makes it important for the client not to be engaged in conversation during a meal which would greatly increase the chances of aspiration.

There is increased movements and coordination of the tongue which impairs their ability to prepare food for swallowing. Food materials are often pushed out of the mouth by involuntary movements and the entire timing of the swallowing sequence may be disrupted which may cause them to swallow large particles of food and increase the chances of choking. With the timing of swallowing off it may cause a person to swallow when the airway is still exposed causing aspiration of food particles which introduces foreign particles into the airway and lungs, causing aspiration pneumonia.

Other factors that affect swallowing is the environment, they need a quiet undistracting eating environment with minimal to no conversation, only the caregiver giving cues to chewing and swallowing. They need to be able to concentrate only on the task at hand of eating and swallowing. If the person with Huntington's Disease is tired or anxious they may have more difficulty with chewing and swallowing.

With anxiety, the muscles of the throat are particularly vulnerable as it gives the sensation of a lump in their throat when they are extremely upset. Thus they have more difficulty trying to coordinate their swallowing as the muscles are more tense and more difficult to try to control. This anxiety can become heightened for a person with Huntington's Disease especially at mealtime if they have had an experience of choking or near choking before.

So it is important to have a plan of how to help your client be relaxed at mealtime and not to fear an upcoming meal. If they have nearly choked or choked in the past remember to use a lot of reassurance with them and help them by giving them the verbal cues of the chewing and swallowing process. This will help them stay focused on the task at hand and may distract them from their anxiety which will aid them in relaxing more. We have a swallowing video available with our display if anyone is interested in it.

Some Warning Signs of A Swallowing Problem – see page 19 for posting in a patients room.

Tips For Mealtime – see page 20 for posting in a patients room.

As with people's moods, swallowing difficulties and changes can fluctuate from day to day or from week to week. This does not mean that with one meal or one day of real difficulty swallowing that a person with Huntington's Disease is progressing to not being able to swallow at all, but it does put them at that time at a higher risk for a choking episode.

Be patient and understanding, give a lot of reassurance and encouragement as it can be a real distressing time for your client. Let them know that they can take a break, maybe they are just too tired or had a more anxious day, encourage them to rest, but also explore with them if there is anything else bothering them. There maybe something else distressing them, pain, anxiety , tired, scared or maybe it could be as simple as they do not like the meal.

Talk to your client and try to find out a problem. If there does not appear to be anything obvious just stop the meal and try something else at the next meal or when they feel hungry again. You may want to try some supplements in between meals such as Ensure to ensure that your client does get their calorie intake.

As the disease does progress you will notice that your client will have more and more difficulty with different foods and textures, keep assessing their swallowing daily and what foods are easier for them.

Choking and aspiration pneumonia are the largest causes of death in persons with Huntington's Disease and can happen quite unexpectantly. When there are more and more problems with eating and swallowing and noticeable weight loss it is time to be in touch with your client's doctor to discuss further strategies.

One issue that does come up as the disease progresses and swallowing becomes more and more difficult and your client begins having episodes of choking or even getting aspiration pneumonia, is the issue of a possible feeding tube. This issue has quite a controversy around it and needs to be discussed with the client, their family and their doctor. It eventually comes down to quality of life issues and is not the right choice for everyone. Some people want it and others do not, but the dependant factor is if it is prolonging your client's suffering or will it give them a prolonged quality of life.

I would now like to share with you a personal experience that I had with my grandmother in relation to eating and swallowing difficulties.

My grandfather opted to keep my grand-mother home and care for her himself and he did a wonderful job. My grandmother would not allow him to have any help with her care besides family members and he promised her he would not put her in a home but care for her until the end.

Well even though my grandfather had taken into account everything basically that I have discussed here today in all aspects of care including feeding, my grandmother got down to a weight of approximately eighty-five pounds. She ate Ensure puddings, baby food custards, cream of wheat and high calorie custards, but she was having a harder time with her swallowing.

One very upsetting and disturbing memory was of trying to feed her myself, with each and every spoonful my grandmother would be coughing and making a gurgling sound. I had to talk her through the swallowing process and she used to always do the double and triple swallow, but it was never completely effective. She would swallow her food and her saliva would run out her nose, each meal we would go through almost a half a box of kleenex for her nose, it was a continuous flow out her nose of saliva.

Seeing my grandmother like this was very upsetting and difficult for me even though I knew that this was how the disease progressed. My family has had much experience over the years with Huntington's Disease and were very open with it, I discussed with my grandfather about thinking it was time to speak to the doctor about a feeding tube. This was our decision with my grandmother knowing that she still could live a quality life with a tube. It was not until two months later that my grandfather had a bad experience at home with my grandmother of choking. This scared both him and my grandmother, she went into hospital and ended up with choking to the point with each meal of needing to be suctioned.

Our decision then came to light, it was time for the feeding tube. Since the feeding tube has been put in last summer my grandmother is a changed woman, though now in a longterm care facility, she has gained a fair amount of weight, now she is over a hundred pounds, and is also brighter. We thought that she was deteriorating cognitively more that she should have been as she was seeing things not really there and did not know all the family. This is not the case now, she is as bright as a whip now. You ask her anything about anyone in the home around her and her family and she will know what is going on.

The cognitive changes we noted as so bad were actually due to starvation as she was not receiving enough nutrition to sustain herself due to her swallowing difficulties. Though with a feeding tube in place a person with Huntington's Disease can still get aspiration pneumonia, this due to aspirating on their own saliva and not being up and around, so even though they get their proper nutrition through their tube feeds and lower their chances of choking and aspiration, it can still occur.

So this again brings up the issue of quality of life even with a feeding tube, so this in itself becomes a controversy.

Dental Care

Oral and dental care are important with persons with Huntington's Disease and as the disease progresses it becomes more of an issue. They have a high calorie diet and have a difficulty clearing their mouths as swallowing becomes more impaired. They have increase in saliva production and a more difficulty swallowing it.

After a meal you may notice food particles remain in the mouth, thus an increase in bacteria stays within the mouth increasing the chance of cavities and increasing the chance of the person swallowing or aspirating saliva with a higher bacteria count. This increases the chance of infection in the mouth and in the lungs, possibly leading to aspiration pneumonia. They also can regurgitate their food which increases the acid in the mouth.

As a person with Huntington's has chorea movements and cognitive impairments it is difficult for them to clean their teeth properly. It is uncomfortable having someone putting a toothbrush in their mouth, and even the use of toothpaste is difficult for the client to spit out as it is so thick. So brushing the teeth is important, but you may need to modify it by using mouthwash instead of toothpaste on the toothbrush or even use toothlette sponges with mouthwash.

It is important to do mouth care after each meal and especially before bedtime to aid in reducing the chance of tooth decay. Always help the person relax prior to doing oral care so it is a more comfortable experience. Their movements increase when they are stressed so the task would be more uncomfortable and stressful for the client and caregiver.

Another issue is that due to chorea movements a person with Huntington's Disease may have an increase in grinding their teeth which too breaks down the tooth enamel and making the teeth more susceptible.

Dental checkups are important for everyone but with persons with Huntington's Disease they should have dental checkups and cleanings done every three months.

Remember to aid the situation by helping with the positioning of your client and helping them be more relaxed so as to decrease the movements at the time of a dental visit.

In relation to dental care issues, it is important to assess your client for tooth and gum pain. These may be brought to a caregivers attention by the client verbalizing pain, but more times than not due to difficulty in communication a client may not be able to verbalize their pain.

So then how do you know they are having pain ? You may notice simple non-verbal cues such as them rubbing their mouth or drooling more, but most important is to investigate any increase in agitated, irritable behaviors. This type of behavior itself is usually always a sign that something is bothering a person with Huntington's Disease, so remember to investigate it, and assess when, where and why their behavior has changed.

Toileting Tips

Due to chorea movements, dystonia and problems with unsteady gait, there needs to be special attention given to the set up of the bathroom.

Most importantly the toilet seat. Persons with Huntington's Disease usually flop down on the toilet, so it is important to have a secure toilet seat and padding on the toilet seat cover and tank will impact the flopping down.

As the disease progresses a raised toilet seat will be needed. With men that stand to void, will have difficulty aiming urine into the toilet so there will be urine on the floor, increasing chances for falls.

With the progression of the disease there becomes a difficulty in completely emptying the bladder as there becomes less control of the muscles of the bladder. As they are drinking more fluids due to increased thirst they have the urge to void more even if they just finished voiding.

Do not remind them that they just voided, do not make them wait as they may have an accident which would cause them embarrassment.

Constipation may become an issue due to a person with Huntington's eats a higher calorie low fiber diet to keep weight on and has a more sedentary lifestyle. More changing in positioning is helpful.

One important issue to remember is to not start using attends too soon. When they are unable to get up on their own, the caregiver needs to assist the person up to the bathroom or commode, or even use a bedpan.

As long as a person with Huntington's Disease has control over their bladder and bowels and still has the urges to void and have a bowel movement, they should not be put in attends unless they request it.

Dressing Tips

As with all activities of daily living it is important not to do everything for a person with Huntington's Disease, this is even the case with dressing. Always allow them to do as much as possible on their own, no matter how minimal it is. By allowing them to do even a minimal task, such as pulling their shirt over their head, allows and helps them feel some independence.

Persons with Huntington's Disease tend to perspire more so their clothes should be changed and washed often. Though they perspire more, which could be related to medications, they more often are more comfortable in a more cooler room, in the winter they may wish to wear thin cotton clothing and even have a fan on them.

This all being related to altered temperature regulation and aspects of the disease related to metabolism. As the disease progresses a small number of people experience recurring high fevers, reaching as high as 40 degrees Celsius at times and even higher.

Many of these fevers occur despite a physicians best effort to identify an infection. So it is important to keep an eye on temperatures in the room and consult a doctor if the client experiences a temperature, so infections can be ruled out or if one present, treated immediately.

They need to have clothes that are easy to put on and take off, loose enough to be accommodating to the chorea movements and withstand frequent washings. Due to difficulty with fine motor tasks, clothing with zippers and buttons are difficult for a person with Huntington's early on, so tracksuits are much easier to maneuver.

Clothing with patterns on them camouflage spills and stains and fashionable patterned scarfs catch drool as effectively as adult bibs.

Contractures

In advanced stages of Huntingtons Disease the ability to control movements is severely compromised.

Even with some involuntary movements present, the client becomes unable to change their own position, maybe more rigid and become at a higher risk of developing contractures.

A contracture is the shortening of a muscle that is permanent and whether with or without pain causes deformity. They occur due to not moving or using the muscles, the tone is lost, thus shortening then occurs.

Contractures become difficult to prevent and manage due to the fluctuations in muscle tone and involuntary movements

Tips to Prevent Falls

Nursing Care In Late Stage Huntington Disease

By Joanne Hunt, RN

Some Tips To Prevent Falls	Environmental Changes to Prevent Falls
<ul style="list-style-type: none">• Do not call out to a client from behind, which would cause them to turn around abruptly and possibly lose their balance• Do not interrupt a person abruptly• Do not give medications while they are standing and do not try to feed them standing up• Do not step in to try to prevent client from "bouncing off the wall" this could actually cause them to fall• Proper footwear is very important. In the later stages high top sneakers or light weight work boots are more likely to be helpful• Most common places for falls are in front of a chair, a toilet and/or a bed• Try "touch-turn-sit" strategy• Orient client to their surroundings and teach about any peculiarities of any of the equipment in their surroundings• Encourage your client not to get up or do transfers without someone present	<p>It is estimated that a person with Huntington's Disease needs twice the space than that of an average individual, thus it is important for no clutter and free of hazards</p> <ul style="list-style-type: none">• Stabilize the furniture so it can not move• Use chairs with armrests and high backs• Clear away any unused furniture, providing clear accessible paths• Remove any throw rugs and thick carpeting• Rearrange tables and lamps away from most frequently used paths• Place padding on frequent bumped furniture and doorways and/or have them rounded off• Side rails on a hospital bed• Tub rails and tub seats• Raised toilet seat

Some Warning Signs of A Swallowing Problem

Nursing Care In Late Stage Huntington Disease

By Joanne Hunt, RN

<ul style="list-style-type: none">• Clearing the throat frequently• Wet or gurgly sounding voice• Coughing while eating and after a meal• A noted delay prior to swallowing food• Not swallowing food and liquids but rather holding them in the mouth• Having difficulty coping with different textures of foods and liquids• Exaggerated movement of the mouth and tongue• When eating and drinking tilting head back	<ul style="list-style-type: none">• When swallowing one bite of food, noted to swallow numerous times• Food and liquid falling out of the mouth or spitting out food• Still having food left in the mouth after swallowing and a completed meal• Having excessive secretions through the nose and/or mouth while eating• Showing signs of fatigue during meals• A noted weight loss over time• Recurrent episodes of pneumonia
--	--

Tips For Mealtime

Nursing Care In Late Stage Huntington Disease

By Joanne Hunt, RN

-
- | | |
|--|---|
| <ul style="list-style-type: none">• Minimize distractions during meals, concentrating mostly on swallowing properly, encourage no talking while trying to eat, save conversation for after meals• Help the client eat until they are full• Make sure they eat slowly, chewing food completely before swallowing• Be careful with hot beverages due to the decreased sensation to heat• Some people find it easier drinking through a straw• Use a single or double handled cup, and even a mug or cup with a cover on it, to help preventing spills, especially of hot liquids• Using a teaspoon can help in smaller amounts of food put into the mouth at a time• Make sure the client is properly positioned
Proper positioning being<ol style="list-style-type: none">1) sitting upright2) swallow with head tilted down with the chin toward the chest3) Find ways through experimentation to minimize head movements | <ul style="list-style-type: none">• Ensure to choose food of appropriate texture and temperature• Soft, blended and pureed foods are generally easier to swallow• Thickened liquids are easier to swallow• Acidic foods increase saliva production so try to avoid while sweet foods decrease saliva production• Make sure your client takes small bites of food• Try to alternate between solid food and liquids• Try to encourage a dry swallow or to take double swallows between each mouthful of foods and liquids• As a caregiver, it is important you take your time as well, do not rush your client when they are eating• Have your client remain sitting up after eating for up to 20 - 30 minutes• Report any coughing or choking incidents to your supervisor and/or doctor• As a caregiver learn the Heimlich maneuver |
|--|---|
-

