Huntington Society of Canada
HUNTINGTON
Société Huntington du Canada

End-of-Life Care in Huntington disease

The World Health Organization defines palliative care as an approach to care that improves the quality of life of patients and their families facing the problems associated with life-threatening illness. This is achieved through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual.

End-of-life care is part of palliative care and is used to describe the support and medical care given during the time surrounding death.

In addition to palliative care, end-of-life care is active care aimed at helping patients and families to:

- Prepare for death
- Ensure comfort
- Make care decisions that are consistent with the patient`s prognosis and goals of care
- Receive bereavement care after the death of a loved one

For the purpose of this fact sheet, the beginning of end-of-life in HD is determined as the stage when the person affected has little control over movement, is bedbound, unable to communicate, unable to eat and drink on his/her own and experiences severe chorea or extreme rigidity.

Caring for someone with HD in the end stages of life should be no different than caring for anyone else at the end of life. Person-centered and holistic care should be provided by a multidisciplinary team that is working in close cooperation with the patient and his/her family with a clear focus on quality of life.

Decisions about end of life preferences will ideally be discussed and documented while the person affected by HD is still capable of making informed decisions and before he/she loses the ability to communicate. These decisions include which medical treatments and interventions the person wants to undergo and which ones he/she would prefer to have withheld (e.g. resuscitation, ventilation, antibiotics, feeding tubes, organ/tissue donation).

Common symptoms at the end-of-life: Long term neurological conditions such as HD can make it challenging to determine if end-of-life is near as disease trajectory is so individual and often difficult to foresee. Common symptoms at the end of life are identified as significant weight loss despite high caloric intake, isolated episodic fevers despite negative blood and urine cultures, respiratory distress secondary to repeated aspiration or infectious pneumonia and sleeping or deep lethargy for most of the 24-hour day.

Causes of death: People die of life-threatening conditions related to HD. The main causes of death are respiratory disease (pneumonia), cardiovascular disease (heart failure, heart attack) and "other HD related causes" (e.g. malnutrition, dehydration, choking).

Providing care for a person in the end stages of HD requires a thorough understanding of the clinical characteristics of the disease - to allow appropriate responses to individual needs, to maintain the highest possible quality of life and to ensure all comfort measures are provided.

What to know when providing care for a person affected by HD who is reaching the end of life:

Altered perceptions: A person affected by HD can experience altered perceptions, most often of pain and temperature. At times, other senses (smell and touch) can be altered. This can cause significant discomfort for the person who may have no means to communicate his/her distress.

Aspiration/choking/infections: Swallowing abilities will become severely impaired. The person with HD can choke on and aspirate not only food particles, liquids and medication but also gastric reflux, saliva and nasal secretions. Feeding tubes will not prevent aspiration or choking incidents.

End-of-Life Care in Huntington disease

Huntington
Society of Canada
151 Frederick St.,
Suite 400,
Kitchener ON
N2H 2M2
1-800-998-7398
info@huntingtonsociety.ca
www.huntingtonsociety.ca
Charitable Registration
Number
11896 5516 RR0001

Contractures: The lack of muscle control, decreased use of muscle groups and changes in muscle tone can lead to severe shortening of muscles. Use frequent position changes and passive range of motion stretching exercises as suggested by the physiotherapist who may also explore the use of soft splints lined with air bladders to support wrists and ankles and prevent flexed posture.

Dental Care: Dental and oral care is an often ignored part of a patient's quality of life. Care needs to be continued and even intensified when the patient has no teeth or is fed though a PEG tube. Consider using an electric toothbrush for effective cleaning.

Feeding tube: A person in the advanced stages of HD will often meet all indications for a feeding tube (e.g. significant weight loss, inadequate hydration, repeated aspiration, severe swallowing incompetence). In the end stage of the disease, however, the person often does not want to eat and drink anymore. This loss of appetite and decrease in thirst is a natural part of the body starting to shut down. The body is unable to use the nutrition, so the decreased intake is not 'starving to death'. Providing nutrition at this stage could actually cause discomfort, and tube feeding might not be appropriate. Instead it can lead to pain, agitation, need for physical restraint and pressure ulcers. Feeding at this stage can prolong suffering instead of increasing quality of life.

If the person with HD and his/her family previously opted for a feeding tube, the need might arise to discuss the discontinuation of tube feeding when reaching the end of life. In some situations, it may be helpful to seek counsel from the ethics board. The overarching goal should always be to maintain the highest possible quality of life for the person suffering from HD.

"Huntington's disguise"(locked-in state of being): A person in the end stages of HD will most often not be able to respond to questions and approaches in a way (either verbally or through gestures) that would be comprehensible. In addition, the person may also lose control of facial expressions and thus will not be able to convey messages about feelings and sensations (e.g. frowning might be missing in a person who is experiencing discomfort). Always assume that the person has comprehension of his/her whereabouts and of the situation, and hears and sees you. It is the ability to communicate that decreases, not the need. Focus on the person while performing caregiving tasks, and keep the person informed about what is happening.

Infections and use of antibiotics: In the end stage of the disease, infections are very common (e.g. pneumonia, urinary tract, skin). While antibiotics may modestly prolong life, seeking treatment may not improve quality of life and may cause suffering. Possible benefits and detrimental effects of antibiotics should be discussed so that informed treatment decisions can be made.

Know the person: Be aware of the preferences and dislikes of the person with HD and provide a sense of control and safety by establishing routines and schedules according to the individual needs of the person.

Pain: If you suspect the person with HD might be experiencing pain, consider providing prophylactic pain medication. Some pain scales offer a checklist of non-verbal pain indicators. When properly dosed, opioids can provide powerful relief for both pain and respiratory distress. Opioids can be safely given at the end-of life without hastening death.

Skin breakdown: Increased attention to skin care, frequent repositioning and specialized seating and bedding as suggested by the occupational therapist will be required as reduced mobility, increased rigidity or chorea, malnutrition, dehydration and weight loss increase the risk of skin breakdown. Keep in mind that although some people with HD appear to be moving all the time, the person might not be able to change his/her position voluntarily.

Spiritual needs: Despite limitations in processing and communicating information, the person with HD can recognize and appreciate familiar rituals, songs, pictures and prayers. It is important to ensure that the individual's preferences and spiritual needs are known and honoured.

Focus on quality of life at the end stage of the disease. Aggressive treatment approaches can lead to a prolonged dying process which may not provide the best quality of life.

Ideal care scenario: The person feels at home, is surrounded by personal possessions and individual preferences are known and honoured. Personalized arrangements have been made for sleeping, seating, hygiene, feeding and daily activities, and needs are reassessed on an ongoing basis. Regular routines are established that create a sense of safety and control, and a consistent team of health professionals cares for the person with HD. Staff receives ongoing education, supervision and support. Family members feel understood and supported.

Visit the Huntington Society of Canada's website (www.huntingtonsociety.ca) to find your local HSC Family Services team member and to access additional information, references, and support.

July 2016