END-OF-LIFE CARE FOR PEOPLE WITH HUNTINGTON DISEASE

End-of-life care is the support and medical care given during the time surrounding death. Its aim is to help individuals with Huntington disease (HD) and their families to:

- Ensure comfort and prepare for death
- Make care decisions in line with their expected outcome and goals for care
- Plan to receive bereavement care after the death of a loved one

It is best to discuss and write down end-of-life preferences while the person with HD is capable of making informed decisions. It is also important to do so before they lose their ability to speak. These decisions include which medical treatments and interventions the person wants (e.g., CPR, breathing tubes, antibiotics, feeding tubes, organ/tissue donation). For more details on this, please see our End-of-Life Options for People with Huntington Disease Fact Sheet here.

End-of-life care includes a:

- Person-centered and well-rounded approach
- Team of medical professionals working closely with the person and their family
- Clear focus on quality of life

What does end-of-life look like in HD?

As HD progresses, more health care providers are needed to help the individual and their support people provide care.

Signs of end-of-life stage in HD can include:

- Having little control over movement, severe chorea and/or extreme rigidity
- Being completely dependent on others for care
- Unable to speak but may still want to communicate
- Unable to eat and drink on their own

Common symptoms at end-of-life

At the end-of-life, common symptoms can include:

- Significant weight loss despite a high caloric diet
- Fevers, even with blood and urine tests showing no signs of infection
- Breathing difficulties, aspirations, and/or pneumonia
- Sleeping or deep lethargy most of the time

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Causes of death in HD

People die of life-threatening conditions related to HD. The main causes of death are pneumonia, heart failure, heart attack, and "other HD-related causes" (e.g., malnutrition, dehydration, injuries from a fall, choking). Some things to keep in mind include:

Aspiration/choking

Problems with swallowing will become much worse. The person with HD may choke on and inhale food, liquids and medication. They can also experience gastric reflux. Saliva and nasal secretions may get into their lungs (aspiration). Feeding tubes will not prevent choking or aspiration.

Feeding tube

Someone at the end-of-life with HD may show signs of needing a feeding tube. For example, the person often will not want to eat and drink anymore. However, 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 reduced intake is not 'starving to death'. Providing nutrition at this stage could actually cause discomfort, and tube feeding may not help to enhance quality of life. It could lead to pain, agitation and pressure ulcers.

If the person with HD had chosen a feeding tube at an earlier time, the discontinuation of tube feeding will need to be discussed at the end-of-life. In some situations, it may be helpful to seek counsel from the facility's ethics board.

Contractures

The lack of muscle control, decreased use of muscle groups and changes in muscle tone can lead to severe shortening of muscles, also called contractures. To help reduce them, try frequent position changes and passive range of motion stretching exercises. The use of soft splints lined with air bladders can help support the wrists and ankles and prevent flexed posture.

Skin breakdown

Attention to skin care, frequent repositioning, and specialized seating and bedding may be required. Reduced mobility, increased rigidity or chorea, malnutrition, dehydration and weight loss increase the risk of skin breakdown. Some people with HD appear to be moving all the time, but may not be able to change their position voluntarily.

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"Huntington's Disguise" (locked-in state of being)

A person in the end stage of HD will not be able to speak or communicate in an understandable way. In addition, the person may lose control of facial expressions. They will not be able to express how they feel or if they have pain. Always assume the person understands, 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.

Altered perceptions

A person affected by HD can experience altered perceptions, most often of pain, temperature, smell and touch. This can cause significant discomfort for the person who may not be able to speak to communicate his/her distress.

Pain

Pain medication may be provided to the person with HD to address suspected pain. Members of the medical team can offer a checklist of non-verbal pain indicators. When properly dosed, medications can provide powerful relief for both pain and respiratory distress and be given safely at the end-of-life without hastening death.

Oral care

For improved quality of life, continue oral care even if the patient has no teeth or is fed though a feeding tube. Consider using an electric toothbrush.

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 prolong life a bit, they may not improve quality of life and could cause suffering. Benefits and side effects of antibiotics should be discussed with the medical team so informed treatment decisions can be made.

Spiritual needs

Despite limitations in communication, the person with HD can recognize and appreciate familiar rituals, songs, pictures and prayers. Ensure the individual's preferences and spiritual needs are known and honored.

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Tips for quality of life at the end stage of HD:

- Surround the person with supportive people and make them feel at home
- Understood and honour personal possessions and preferences
- Establish and reassess plans for sleeping, seating, hygiene, feeding, daily activities, environment
- Use routines to create a sense of comfort and control
- Ensure family members feel understood and supported

For more info, please see HSC factsheets on End-of-Life Options for People with Huntington Disease and Feeding Tubes in HD, or the Physician's Guide and Carer's Guide.

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