Huntington's Disease Association

End of life care for people living with Huntington's disease

EUROPEAN HUNTINGTON'S DISEASE NETWORK

Advancing Research, Conducting Trials, Improving Care

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Contents:

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Community reviewer

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Introduction

This guideline sets out best practice in end of life care for people living with Huntington's disease.

During the final stage of the illness, the person is no longer able to work or manage their own finances, personal care and domestic responsibilities. They will also have difficulty with mobility, needing to be in a chair or bed for the majority of the time. At this stage, swallowing may be problematic or difficult and there may be significant weight loss. This stage may last for many years, especially if the person is fed by artificial means. Quality of life may appear to be very limited and families may find this an emotionally draining time and need extra support from healthcare professionals.

In the advanced stages of the disease, people will find all communication very difficult and may choke on food and experience lung infections.

Planning for end of life

Offer the person the opportunity to discuss their preferences and concerns about care at the end of life at trigger points such as at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and consider the person's current communication ability, cognitive status and mental capacity. Consider that the person has probably witnessed, and possibly cared for, other family members with the disease.

Be prepared to discuss end of life issues whenever people wish to do so.

Provide support and advice on advance care planning for end of life. Topics to discuss may include:

- What could happen at the end of life, for example, how death may occur
- Providing anticipatory medicines in the home
- Advance care planning, including Advance Decisions to Refuse Treatment (ADRT) and Do Not Attempt Resuscitation (DNACPR) orders, and Lasting Power of Attorney. Offer people the opportunity to talk about and review any existing ADRT, DNACPR orders and Lasting Power of Attorney when interventions such as gastrostomy and non-invasive ventilation are planned
- Ensuring advance care plans will be available when needed, for example, by including the information on the person's Summary Care Record
- When to involve specialist palliative care

Areas that people might wish to plan for, such as:

- What they want to happen (for example, their preferred place of death)
- What they do not want to happen (for example, being admitted to hospital)
- Who will represent their decisions, if necessary
- What should happen if they develop another illness

Think about discussing advance care planning with people at an earlier opportunity, given the impact of Huntington's disease on communication ability, cognitive status and mental capacity.

Provide additional support as the end of life approaches, for example, additional social or nursing care to enable family members to reduce their carer responsibilities and spend time with the person with Huntington's disease.

Towards the end of life, ensure there is prompt access to the following, if not already provided:

- A method of communication that meets the person's needs, such as an Augmentative and Alternative Communication (AAC) system

- Specialist palliative care
- Equipment, if needed, such as syringe drivers, suction machines, riser–recliner chair, hospital bed, commode and hoist
- Anticipatory medicines, including opioids and benzodiazepines to treat breathlessness, and antimuscarinic medicines to treat problematic saliva and respiratory secretions. Opioids should only be prescribed under the supervision of a palliative care or pain specialist

Offer bereavement support to caregivers (as appropriate)

Quality of life

It can be difficult when someone is in the advanced stages of Huntington's disease to think of activities and things to do to interact with them.

It can also be easy to assume, because of their lack of verbal and non-verbal communication, that the person does not understand what is happening around them. This is not the case, and the person can still hear, see and understand. It is therefore important to try to engage with the person.

Be aware of the need for continued stimulation. People with Huntington's disease still have hobbies and interests, and it is essential that they can pursue these. If possible, refer to an activity co-ordinator to assist with this.

Plan activities around their interests and take into consideration their shorter concentration span and the tendency to fatigue.

Consider using tools such as life storybooks, and gather information from family and friends about the person's hobbies and interests. This will help target activities that the person may want to engage with.

Be aware that people with Huntington's disease will still have sexual needs. Help ensure they are in an environment that will enable them to facilitate this. If there are concerns about the person's mental capacity to consent to sex, then ensure this is assessed. Other forms of intimacy between the person and their loved ones, such as touching and cuddling, also remain important as the disease progresses.

Looking after someone's personal hygiene in advanced stage Huntington's disease can be difficult. Cognitive changes can make a person uncooperative, and physical symptoms may present some challenges.

Creating the right environment, which is warm, secure, structured and calm, will make the person less anxious and insecure.

Having a daily routine and talking through this with the person can be helpful.

Oral hygiene and dental care are important when someone is at an advanced stage of Huntington's disease. Consider the following steps to help promote this:

- Ensuring that someone's mouth is empty following a meal
- Cleaning teeth regularly with a specially adapted brush, such as 'Dr Barman's Super Brush'
- Referring the person to a community dentist for regular check-ups, if visiting the surgery becomes difficult. Weight loss may occur if ill-fitting dentures are worn, so these should be checked at regular intervals

There are several reasons why people with advanced stage Huntington's disease are more susceptible to the development of pressure sores. These include weight loss, poor diet, involuntary movements and / or rigidity and stiffness.

It is difficult to heal a pressure sore once formed so vigilance and regular pressure area care is essential.

Consider a referral to a physiotherapist and occupational therapist to help with the provision of appropriate equipment and positioning of the person. Chest physiotherapy can be helpful in advanced stage Huntington's disease to reduce the risk of aspiration pneumonia.

There are multiple professionals involved in the care of someone with advanced stage Huntington's disease and this can generate many different hospital appointments.

Evaluating which appointments are necessary and carrying out some updates on the person's condition by phone may reduce the need for some of these appointments.

Gather as much relevant information as possible from family and friends about the person. It is important to know if there are any significant people (or animals) that form an important part of their life.

Establishing any goals or priorities and helping the person achieve these can influence their quality of life.

Having an awareness of someone's religious and spiritual needs, and helping these be met, can provide comfort and reassurance.

An awareness of religious and cultural needs leading up to and immediately after death may be particularly important in some religions so staff need to be aware of these and ensure they are followed.

Caring for someone with Huntington's disease involves the care of the whole family. Unpaid carers should seek a carer's assessment. People with advanced stage Huntington's disease can be particularly emotionally challenging.

Communication with a person with Huntington's disease

As Huntington's disease progresses, the ability to communicate effectively decreases.

People who have the disease will develop dysarthria (interruption in muscle control, which affects speech) and chorea movements will make speech difficult to understand. Capacity to make decisions is often impaired.

Taking the time to develop a personal-professional relationship is vital to effective communication with someone who has Huntington's disease. If this is challenging to do because of the person's communication difficulties, then ask caregivers about the person's likes and dislikes.

Cognitive changes may have a profound effect on organisational skills, and a person may demonstrate impulsivity, lack of insight and difficulties in learning new information. They may also develop perseveration (when a person is "stuck" on a particular thought or action and struggles to disengage from it). For example, a person could perseverate regarding a particular item, such as bottles of water, and buy it in excess. They may also ask questions on the same subject repeatedly, which can be a source of anxiety for the person and stressful for those supporting them.

This can make it difficult to communicate with the person effectively.

A person with advanced Huntington's disease is likely to still have memories of their life before developing the disease and the ability to comprehend what is happening, even if their communication skills have decreased.

Seek advice from caregivers on how to communicate effectively with the person.

Request a comprehensive ongoing assessment of communication skills from a speech and language therapist. They can make recommendations about communication aids that can help. This can include equipment, such as communication charts, books and apps. Some of the more technical communication aids may not be suitable for people who are having problems with manual dexterity or who may have issues learning how to use these aids. Be aware that a decline in cognition may prevent a person from developing the skills to effectively use a system, particularly if it is complex.

A communication aid is most effective when used regularly. A referral to a speech and language therapist can be made via a GP or a specialist Huntington's clinic.

Be prepared to adapt communication techniques to ensure effective communication can take place.

Break down sentences into short parts and do not repeat the question too soon if a person does not reply immediately – give them extra time.

Time and patience when communicating is important, as is reducing the amount of background noise, because this can be disturbing to the person's cognitive processes.

Look for non-verbal clues and monitor for change in sound of any speech that the person is able to make.

In the advanced stages, the ability to draw upon previous experience with the person will assist with the assessment of symptoms.

Ensure that if a person has decreased communication ability, that there is a method in place to communicate their information between professionals.

Symptom management

Effective management of symptoms in advanced Huntington's disease is crucial for the person and their loved ones. However, the disease complexity and duration of this phase of the condition can make this challenging.

A multi-disciplinary team approach is needed to meet the needs of people who have advanced Huntington's disease. Neurologists, psychiatrists, psychologists, physiotherapists, occupational therapists, speech and language therapists, dietitians, nursing and social workers should all be involved in providing assessment and treatment.

If a person's symptoms change or deteriorate, then explore why this has happened. For example, does the person have an infection or are they in physical pain? Are they feeling distressed, frustrated or anxious because of a change to their routine or environment?

Pain

People with advanced Huntington's disease can experience pain for many reasons.

As with other symptoms, it can be difficult to assess this when a person is unable to communicate and / or is cognitively impaired. However, a pain assessment should try to determine the likely cause of pain so that the approach to management is appropriately targeted.

The use of validated pain scales for people who are non-communicative and / or have dementia, such as the Abbey Pain Scale (Abbey 2004) can be helpful.

Seek advice from palliative care specialists on the use of such tools.

Spasticity, rigidity, stiffness and myoclonus can cause pain and should be treated accordingly.

Consider non-pharmacological interventions, such as physiotherapy and postural management.

People with Huntington's can experience neuropathic pain (persistent pain caused by problems with the nervous system).

If a person is experiencing neuropathic pain, seek advice from a neurologist or pain expert on appropriate imaging. The necessity and potential benefits of this should be weighed up along with the risk of distress to the person.

Consider whether neuropathic pain can be managed with medications, such as gabapentin, pregabalin and amitriptyline.

Consider non-opioid analgesics like paracetamol before considering opioid analgesics like co-codamol and Oramorph. Referral to a palliative care or pain specialist should be offered if pain is not being effectively managed.

Be aware that people with advanced Huntington's disease can experience pain for reasons that are not directly related to their condition. For example, dental problems resulting in dental pain are common in people with Huntington's disease.

Discomfort caused by constipation and urinary tract infections can also occur and should be treated accordingly.

Toileting

In the advanced stages of Huntington's disease, people usually become incontinent of both urine and faeces.

Ensure that appropriate continence products are used.

Regular pads may be ineffective, for example, if a person has chorea. If this is the case, then seek advice from an NHS Continence Service.

If a person requires catheterisation, then consider using a suprapubic catheter. This can be the best option if a person requires a catheter for long-term use because people can find them more comfortable and easier to manage.

People who are still continent often experience urinary frequency and urgency, which can necessitate frequent visits to the toilet. It is important to exclude urinary tract infection and constipation when this is a new problem. In these circumstances, consider using antimuscarinic drugs such as oxybutynin and tolterodine. However, in men, other causes such as prostate problems should be ruled out first, and potential side effects of medications should be considered.

Urinary tract infections can regularly occur in people with advanced Huntington's disease and lead to an acute exacerbation of Huntington's disease symptoms. Therefore, urgent treatment is important.

Keeping people adequately hydrated can be difficult when they have impaired swallow and no PEG (Percutaneous Endoscopic Gastrostomy) feeding tube.

Constipation is also a common problem in advanced Huntington's disease due to progression of the disease, increasing immobility and poor nutrition and hydration status.

The side effects of medications can also contribute to constipation. Manage constipation with appropriate laxatives and by increasing fluids and fibre in the diet, where possible.

Temperature dysregulation

It is not uncommon for people with advanced Huntington's disease to feel very hot and to experience episodes of profuse sweating and flushing.

Rule out other potential causes for this such as infection, medication side effects or hormonal problems.

The feelings of heat can be very intense, uncomfortable and distressing. It can also lead to worsening of movement symptoms and agitation.

Consider the following strategies to help manage this:

- Keeping surroundings, such as bedrooms, at a lower temperature, using fans, placing cool damp cloths on the person's forehead / neck or using icepacks wrapped in a towel or cloth
- Ensuring clothing is light and made of natural fibres
- Placing t-shirts in a plastic bag in a freezer to cool them down before the person puts them on
- Giving the person cold drinks and ice cream when they are feeling very hot
- Giving extra water via a feeding tube
- Massaging levomenthol cream into the person's arms / legs, which can have a cooling effect

In the advanced stages of Huntington's disease, people can experience unexplained recurring high fevers where no infective cause is identified. If this occurs, then ensure where possible that the person is hydrated. Consider paracetamol to reduce the fever, as well as using the strategies above.

Hypersalivation

Many people with advanced Huntington's disease experience problems managing saliva and drooling.

This can be distressing for them and cause difficulties if they have impaired swallow as saliva can pool in the mouth and drooling can result in sores to the mouth and chin. It can also make eating and drinking more difficult.

Consider non-pharmacological interventions including postural management and supports for the head and neck and manually assisted cough techniques.

Consider medications such as transdermal hyoscine (scopoderm patches), sublingual atropine, glycopyrrolate, and ipratropium bromide inhalers. Seek advice from palliative care specialists who can advise on these medications and appropriate doses.

Ensure the person is closely monitored to ensure that their mouth does not become too dry and that other mucous membranes and skin do not become dry.

Consider a referral to a specialist botox clinic in cases where medication is not effective.

For people who have thick secretions, ensure that they are well hydrated. Humidifiers, steam inhalation and nebulisers can help achieve this. Mucolytics like Carbocisteine may be helpful.

Nausea and vomiting

Whether a person is still eating and drinking orally or using alternative methods such as PEG feeding, nausea and vomiting can be a problem for people with advanced Huntington's disease.

In the first instance, other possible causes of this should be investigated.

Consider the following questions:

- Is the person constipated or do they have an infection?
- Are they taking any medications which could be causing nausea as a side and are medications being given appropriately to minimise such side effects i.e. with or without food?
- Could medications be contributing to gastro oesophageal reflux?
- Do they have any other symptoms such as diarrhoea, high fever and stomach pain?

Once infection, medication and other causes have been excluded, review when the nausea and vomiting occur. Does it occur directly after eating / drinking? If so, review positioning and seating when the person is eating / drinking.

If the person is still eating orally, consider referring them for a speech and language therapy review.

A person may eat too quickly due to hunger and impulsivity and therefore cram food into their mouths. In these circumstances, advise caregivers to remind them to eat slowly, taking one mouthful at a time.

The person may also be taking large gulps of fluids whilst eating, making them feel sick. They may not be able to chew food and so are swallowing things whole. Having regular small meals rather than three big meals a day can sometimes help and reviewing consistency of foods and fluids is important.

If the person has a feeding tube and is experiencing nausea and vomiting, a review of posture during feeds is important. Consider the following questions:

- If the person has overnight feeding via a pump, are they lying flat?
- Could the feed rate be too quick?
- Is the feed suitable?

Seek advice from a dietitian on adjusting feeding times, rates, volumes and feed types to try and minimise any feelings of nausea.

If the person is experiencing reflux, consider a proton pump inhibitor such as omeprazole or lansoprazole.

Consider antiemetic therapy.

Seizures

It is not uncommon for children / young people with Juvenile Huntington's disease to experience seizures, which should be managed according to standard epilepsy treatment protocols.

Occasionally those in the advanced stages of adult-onset Huntington's disease can have seizures.

Be aware that myoclonic jerks can be mistaken for seizure activity.

Seek an urgent neurological opinion if there are any concerns that someone has had a first seizure. They will carry out appropriate investigations and develop a treatment plan, if necessary.

Last days of life

The most common cause of death in advanced Huntington's disease is pneumonia or infection. Depending on the conversations around priorities and advance care planning, this could be managed in the person's usual place of care, with a focus on comfort and symptom management. Medications to manage symptoms at the end of life can be given via a continuous subcutaneous infusion; specialist palliative care advice should be sought if there is uncertainty around symptom management.

Support following death

Offer family members support following the death of the person. They could seek counselling and access bereavement support through organisations such as Cruse, or contact the Huntington's Disease Association for someone to speak to and advise on organisations that can offer specific bereavement support.

For professionals involved in providing care, it is vital they are self-aware and seek support from other colleagues and professionals. Being part of a team and using staff supervision can also help.

Resources

Huntington's Disease Association resources for professionals

The Huntington's Disease Association offers a range of resources to support professionals working with people affected by Huntington's disease.

You can find our resources at **www.hda.org.uk**

Explore the professionals webpage using the search bar to find specific resources. Search "resources".

Stay informed

Join our professional-only mailing list to receive updates on events, webinars, and new resources tailored to your needs. You can sign-up at **https://www.hda.org.uk/professionals-and-training/**

Support for children and young people

Find information on our Huntington's Disease Youth Engagement Service (HDYES) by searching for "HDYES" on the website. Or visit **www.hda.org.uk/children-and-young-people/**

Other resources

Abbey Pain Scale. Available here.

ACP support. Training in advance care planning. Available here.

General Medical Council. Clinically assisted nutrition and hydration (Treatment and care towards the end of life: good practice in decision making). Available here.

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Get in touch

For advice and support or to speak to a Specialist Huntington's disease Adviser

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