



Huntington's
Disease
Association

**Care in advanced
Huntington's
disease**

We'll be there

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Foreword

“You matter because you are you, and you matter to the end of your life. We will do all we can not only to help you die peacefully, but also to live until you die.”

DAME CICELY SAUNDERS

Caring for someone with advanced Huntington’s disease is both a privilege and a challenge. The hereditary nature of the illness offers another dimension as family care givers are often at risk of developing the illness – so the care we offer their loved ones will or may set the scene as to what to expect.

I hope that you will find this booklet useful and must express my thanks to Bill Crowder, Ruth Abuzaid, Kathy Mardle-Aylett, Rachel Taylor, Desiree Salanio and Alison Heavey and the several family members who have looked at and contributed to the booklet.

Cath Stanley

Chief Executive, Huntington’s Disease Association

Families



Stages of Huntington's disease

Having a family member with Huntington's disease is never easy, but seeing someone in the last phase of the condition can be especially emotionally challenging. It is generally accepted that there are five broad stages to the course of the disease, the final stage being when the person with Huntington's disease is no longer able to work, 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 have been 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 the families may find this an emotionally draining time with some tough decisions to be made and they may need extra support from healthcare professionals.

Difficult conversations

When advising families who are facing the advanced stages of Huntington's disease, care must be taken to give honest advice with sensitivity. Families will need to know of the practical issues they are likely to face and should be prepared to make some difficult decisions, so it is vital they know the facts.

It is by no means inevitable that care at this advanced stage must be provided in a residential or nursing home. Depending on the accommodation, many people are able to stay at home with a care package provided by Social Care or the NHS until the end of life. It is ideal if a conversation about where care should be provided is held when the person with Huntington's disease is able to express their views and discuss it, but beginning conversations such as this is not easy, especially if the person with Huntington's disease does not wish to look too far ahead. It is sometimes the case that families make promises about the place of future care, promising never to relinquish their care to a home or hospice. This may lead to difficulties in the future, as it

is not known what circumstances may lead to a need to break that promise, such as the health of the family carer, or a change of home circumstances. This should be borne in mind when discussing future provision of care with family members.

Another issue that families find difficult to broach with loved ones is what they wish to happen when swallowing becomes so difficult they are unable to eat enough to maintain their weight. Ideally, the issue of artificial feeding should be discussed with a healthcare professional such as the Speech and Language Therapist, the Dietitian or one of our Huntington's Disease Association Specialist HD Advisers well in advance of a decision needing to be made. However, not wanting to plan too far ahead is often an issue for people with Huntington's disease and the topic may be avoided. If a Lasting Power of Attorney (LPOA) for Health and Welfare is made, the decision may fall to the attorney(s). If no LPOA or Advance Decision to Refuse Treatment (ADRT) is in place and the person with Huntington's disease lacks the mental capacity to make the decision themselves, a best interest decision by the multi-disciplinary team will be made. It is good practice to ask the family of their preferences and any preferences the person expressed previously, but ultimately it will be the decision of the healthcare professionals.

When care is being provided in a care home setting, staff will usually want to engage the person with Huntington's disease and/or the family in decisions regarding the preferred place of death and whether they wish to have cardio pulmonary resuscitation if their heart or breathing should stop. Many people have a strong desire to end their life in their own bed rather than in a hospital and this should be discussed and noted in care plans. Cardio pulmonary resuscitation is an invasive process and can lead to physical damage such as broken ribs. It may not be the peaceful end to life that most of us hope for, especially if at the end of a long and arduous illness such as Huntington's disease.

Another option that may be available is care by a hospice in the final few weeks either as an inpatient or via a home visiting team. A referral can be made to the hospice team by the GP or doctor caring for the person with Huntington's disease. This may offer a calm and well managed time at the end of life and provides support for the family.

It should be remembered that when preparing a family for the imminent death of their loved one, most people have never seen a person close to, or just after death. They may have no idea of what to expect and be fearful they won't cope. A caregiver should try to not

let their own fears of talking about dying, death and bereavement get in the way of conversations. Some family members have reported a reticence on health professional's part to discuss imminent death and they would have found it helpful to have someone who was open, honest and prepared to discuss what to expect. Despite the long and protracted nature of the illness, many family members are ill prepared to face the final few hours when they arrive and are shocked to be told death may be imminent.

Beginning those difficult conversations is not always easy, but planning for the future and speaking openly can improve a person's quality of life, now and in the future. It can reduce anxiety if a person knows there is a clear plan for the future.

Grief

Grief is not only a reaction to death, it is the natural reaction to any loss including a home, country or relationship. Grief may occur when there is a realisation that there is something you will never have again. This may happen in the later stages of Huntington's disease as loved ones begin to realise there are elements of their relationship that can never be regained. Some families feel that they lost their loved one long before the actual death of the person and that they have already had a long period of bereavement by the time the person with Huntington's disease finally passes away. Many family carers have expressed relief at the point of death and a feeling that they have already done all their grieving and are ready to move on. This may also trigger feelings of guilt as the bereaved feel they are not mourning in the expected way.

Anger after a death of a loved one is a normal and healthy reaction. Some may feel angry at the deceased person – "how dare he leave me?", sometimes it is directed at their God – "how could He allow this to happen?" and sometimes it is directed at the doctors and healthcare professionals involved with the deceased at the end – "they should have done more." These thoughts may help to channel the anger and are a natural consequence of the grieving process and should subside in time.

Families should be given every care and understanding at this stage by the healthcare professionals around them. They may not feel ready to clear away possessions for some time and in a care home setting, delicacy may be needed to negotiate a time scale for a room to be cleared and possessions collected.

Talking to children

When there are young children in a family, it is natural to try and protect them from upsetting discussions and events. Families will often avoid talking about Huntington's disease in front of children, often until they are adults. However, it is important to include children in the person with Huntington's disease's life as much as possible, especially if they are their parent. Discussions should be tactful and information given age appropriate. Parents should always be consulted and agree with any information or planning a child is involved with.

The Huntington's Disease Youth Organisation (HDYO) website has age appropriate information for young children to young adults - see website details on the next page. Children may have specific concerns or questions about genetics. HDYO have some excellent information but also clinical genetic departments will see and talk to children about their at risk status.

Explaining death to children is something most parents worry about and find difficult. When bereavement occurs, we should remember that a child will experience the same range of emotions that adults do, from feelings of shock and disbelief to numbness, despair, anger and guilt.

Children can be very absorbed by their grief but at other times seem to carry on life as normal. They may feel mixed up and confused about issues they do not understand, but feel wary of asking questions, especially if they think their questions may upset grown-ups. Their feelings are as important as any other member of the family and it may be helpful to direct the adults to resources that may help them to manage a child's feelings.

Depending on the age of the child, information and books are available to explain death and explore the feelings a child may have when a member of their family dies.

At the Huntington's Disease Association, we have a team of Specialist Youth Workers who will be able to offer support to young people affected by loss and bereavement.

Support for the family

Families will require more support as the disease progresses and care needs increase. If the person with Huntington's disease is being cared for at home, local hospice services and palliative care teams may be able to offer the family anticipatory grief counselling and advice on

end of life care. Our Specialist HD Advisers at the Huntington's Disease Association may have known the family for some time and are often well placed to offer practical and emotional advice to the family. A family member may not be aware of how near to death a relative is and therefore may not be anticipating it.

The Huntington's Disease Association's branches and support groups, which are run across the country, offer which are run across the country offer peer support and many families take comfort from the experiences and support of others affected by Huntington's disease. A list of our branches and support groups can be found on our website www.hda.org.uk

There is also an online HDA Message Board, where family members can post anonymously their thoughts and feelings and discuss issues with other families affected by Huntington's disease across the UK and the world hdmessageboard.com

After the death, what next?

Families may be so taken up by the day to day coping when a loved one is near to death, that after that death occurs they may feel lost and unsure what to do. They may need guidance from professionals as to what steps need to be taken.

Once the funeral is over, the family need time to adjust and may need to access grief counselling. It is important to remember, that due to the inherited nature of Huntington's disease, the death of one person does not necessarily mean an end to Huntington's disease for a family as there may be other members with the disease or at risk of developing the disease. Families may sometimes have disagreements as to the best way to deal with Huntington's disease and this can lead to strained relationships.


If someone has been caring for a person for many years, their death may leave the carer unsure of their role. They may have given up work to care, and now face having to work again having been out of the workplace for many years. This can seem very daunting. They may have financial difficulties, or need to leave their existing accommodation.


Families may feel that the Huntington's Disease Association's branches and support groups are a source of comfort and advice as many members of the group have faced similar situations already.

References and further reading

Advice can be obtained from:

Bereavement Advice Centre

 www.bereavementadvice.org

 0800 634 9494

Publications for teens and younger children include:


Healing your grieving heart for teens: 100 Practical ideas – Alan D. Wolfelt, Ph.D, 2001


Muddles, Puddles and Sunshine: your activity book to help when someone has died – Diana Crossley, 2000

Useful websites and articles


 www.winstonswish.org.uk

 www.cruse.org.uk

 www.youngminds.org.uk

 www.griefencounter.org.uk

 en.hdyo.org/eve/articles/209

 www.hdyo.org



Communication

Importance of communication

“You matter because you are you, and you matter to the end of your life. We will do all we can not only to help you die peacefully, but also to live until you die.” Dame Cicely Saunders

This quote is the starting point of any communication undertaken with a person with a life limiting disease.

Communication between patients and healthcare professionals is a key aspect to ensure that a trusting and therapeutic relationship exists, as a disease progresses and a person nears the end of life. Effective and open communication allows the person and their loved ones to have a positive experience.

Dame Cicely Saunders, founder of the Hospice Movement, suggested that how people die remains in the memory of those who are left behind (2008). This remains the very real experience of many people who find themselves facing the diagnosis of a life limiting disease, and those who remain after that person has died. How the person and the people significant to them are communicated with is an integral part of creating this memory.

‘More Care Less Pathway’ was a report produced following the withdrawal of the use of the Liverpool Care Pathway. In this report there is a key point identified about communication:

“Unless everyone in society- members of the public, the press, clinicians, public figures- is prepared to talk openly and honestly about dying, death and bereavement, accepting these as a normal part of life, the quality of care and the range of services for the dying, their relatives and carers will remain inconsistent.”

Baroness Neuberger

The report goes on to suggest that there is a requirement for continued training and support for healthcare professionals in ensuring that they have excellent communication skills. Knowledge and technical skill is not enough.

The Leadership Alliance for the Care of Dying People were given the remit of producing priorities of care, based on the recommendations from the Neuberger Report and,

“these make the dying person themselves the focus of care in the last few days and hours of life and exemplify the high-level outcomes that must be delivered for every dying person. The way in which the priorities for care are achieved will vary, to reflect the needs and preferences of the dying person and the setting in which they are being cared for.”

Priorities of care

Five priorities of care have been presented in the report produced by the Leadership Alliance:

One chance to get it right

Priority 1

The possibility that a person may die within the next few days or hours is recognised and communicated clearly, decisions made and actions taken in accordance with the person's needs and wishes, and these are regularly reviewed and decisions revised accordingly.

Priority 2

Sensitive communication takes place between staff and the dying person, and those identified as important to them.

Priority 3

The dying person, and those identified as important to them, are involved in decisions about treatment and care to the extent that the dying person wants.

Priority 4

The needs of families and others identified as important to the dying person are actively explored, respected and met as far as possible.

Priority 5


An individual plan of care, which includes food and drink, symptom

control and psychological, social and spiritual support, is agreed, co-ordinated and delivered with compassion.

Throughout these priorities of care, communication again remains a key component of effective, quality and safe care for an individual.

Importance of communication

How does this equate in practice?

All healthcare professionals, who are caring for those with a life limiting disease, have the opportunity to attend or at least read the literature around these areas, in order to develop their communication skills to be able to initiate open conversations with those with a life limiting disease. Many Trusts offer Advanced Communication Training for Professionals and was the recommendation in The National Cancer Plan  webarchive.nationalarchives.gov.uk

Sage & Thyme, SPIKES (Baile & Buckman) are also examples of communication models which give professionals key points in having difficult, but much needed conversations and a structure for breaking bad news. It is vitally important that healthcare professionals give as much importance to developing their communication skills as they do their practical skills. Excellent communication is the key to providing good end of life care for all.

Important conversations do not have to be based around specific treatments; they should also embrace other areas of a person's life.

Advanced care discussions should embrace what is important in someone's life. When working with those who may have reduced communication, it is also key to understand what their likes and dislikes are for example; fluids and nutrition, style of dress, music and media tastes. This is vitally important; for example the thought of being given something to eat or drink when it is a food or drink that you have always disliked.

The person with Huntington's disease may not wish to have frequent conversation. Professionals should embrace that important conversations can be initiated on frequent occasions; however they have to be led by the person who has the disease. Not everyone wishes to have these conversations, and this should be recognised as a person's right. The professional should also consider that because the person has chosen once not to discuss end of life care, this should not indicate that the person should never be offered this conversation again. Declining

once is not declining forever and is part of having open dialogue with the person. There may be particular triggers in a person's disease progression that can be used as a cue to initiate a conversation.

The professional who is initiating the conversation must show interest and a human element to the subject matter and be able to respond appropriately to the dialogue that they are having. Preparation for important conversations is essential, as is active listening. The environment where this communication is taking place is vitally important, particularly if it is difficult news being delivered. The corridor in a busy hospital ward is not an appropriate place to conduct significant conversation.

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 controlling speech) and chorea movements will make speech difficult to understand. Capacity to make decisions may well be impaired.

In light of this, it is vital that Advanced Care Plans and significant conversations are discussed while a person is still able to reliably perform this and have the capacity to make decisions particularly around areas such as Advanced Decisions to Refuse Treatment (see preparing for the future section).

The previous development of a person-professional relationship is vital to effective communication. Research shows that as the disease progresses, the person may have difficulty in comprehension and the inclination to initiate conversation may decline (Saldert et al 2010).

Cognitive changes may have a profound effect on organisation skills, and a person may demonstrate impulsivity, lack of insight, difficulties in learning new information and develop perseveration, which all have an extreme impact on the ability to communicate with a person effectively. Research suggests that in the later stages of the disease, although the ability to communicate will be severely affected, the ability to comprehend what is happening is still intact. It is vitally important for any professional working with people with the disease that they understand this set of complex behaviours in order that they can adapt their own practice accordingly and will often have to develop ingenious and different ways to communicate with that person.

It is the previous communication and the relationship that has developed with that person that will have a positive benefit on the effectiveness of communication and increase the ability for the person with the disease to participate in daily life. Equally important is exploring family knowledge and using this in a timely and useful fashion. Many families will have had experience of someone else in their family with the disease and this can be an extremely useful resource in that they may have developed skills already, particularly in relation to communication.

Early referral to Speech and Language Therapy is vital, a person with Huntington's disease may be known to a service for the length of their disease. They will be able to provide a comprehensive ongoing assessment of communication skills and provide suggestions of augmentative and alternative communication. This describes the symbols, devices, strategies and techniques that are used to compensate for a person's difficulty with speech, language and communication (Beukelman et al 2005). These can be either high or low tech.

Equipment that has proved to be effective include LightWriter, memory boards, diaries, photo albums, picture symbols, speech boards (Talking Mat) and developing single answer responses through the use of thumbs up and thumbs down. These have to be adapted to allow for the chorea movement or rigidity that may be present for that person.

It is the responsibility of the person communicating with the person with Huntington's disease to take responsibility for their part in communication and to be extremely mindful of adapting their communication techniques to ensure effective communication can take place. Again this demonstrates the need for early development of the person-professional relationship. It is vital to break down sentences into short parts, attempting not to have two parts to a question and never continually repeat the question if a person doesn't reply. Think of the analogy of a clock was given, the hands are at 12, each time a person with Huntington's is asked a question the clock starts ticking. If the question is asked repeatedly the hands keep returning to 12 and the person has to start thinking about their answer again. It is advisable to speak slowly and clearly, however don't treat the person as if they don't have intelligence. A person with Huntington's disease will still have memories of their life before developing the disease and still has comprehension, even if their communication skills have decreased.

Time and patience when communicating is important, as is reducing the amount of background noise that can be disturbing to the cognitive process that is occurring. Equally observing for non-verbal clues, and monitoring for change in sound of any speech that that person is able to make. In later stages, the ability to draw upon previous experience with that person will assist with the assessment of symptoms, or even if they are experiencing hunger or thirst.

It is also important to understand that attempting to introduce new skills to enable effective communication in the later stages of the disease is not advisable, as decline in cognition may prevent a person from developing the skills to effectively use a system, particularly if it is complex.

If a person has decreased communication ability, it is vitally important that there is some method in place to communicate their information between professionals. An area that has been developed in the Dementia community is Life Books, which tells that person's story, however also tells the story of what is important to them now. This is similar to a Patient Passport which some trusts use to enable people with Huntington's disease to have all of the information about their care in one place. The Patient Passport is divided into three sections:



Red

Important information about the individual i.e. personal details, risks, medication including how it should be administered.



Amber

Important information that professionals should be aware of about daily life i.e. communication, sight, hearing, impaired swallow, sleep, behaviour



Green

Important information about likes, dislikes, nutritional/fluid intake etc.

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Preparing for the future

What it means

The diagnosis of Huntington's disease and the experience of living with it have strong repercussions not only for the person affected but also for their families because of its hereditary nature and long duration. The disease is unpredictable and complex hence there are a lot of opportunities to prepare and plan for future care.

Preparing for the future involves sensitive discussion and planning on how best to manage care. Care planning is viewed in the context of an anticipated fatal impact of the disease which brings about the deterioration in a person's level of cognition, subsequent loss of mental capacity to make decisions and their ability to communicate wishes to others.

Importance of thinking ahead

Healthcare professionals can be placed in a difficult situation when a medical crisis occurs because of lack of advance care planning. Important discussions are often not initiated because of perceived fears that they may cause anxiety, hopelessness or that they may create more emotional upheaval.

On the contrary, planning ahead is important as this gives the person, the family and those involved in the care, the opportunity to think about, talk about and document hopes and priorities as the disease progresses in its advanced stage and towards the end of life. It ensures timely access to safe, effective care and continuity in its delivery to meet the person's needs.

Advance Care Planning

Advance Care Planning (ACP) is a process that provides a mechanism for discussion about views, wishes, preferences for future care between a person and their care providers (DH 2009). This gives them the

opportunity to prepare for possible end of life situations which may include discussions of their understanding about their medical condition and prognosis, their personal goals of care, their wishes, values, concerns and personal and family resources.

 www.nhs.co.uk/condition/end-of-life-care

Initiating the conversation

Discussions on Advance Care Planning (ACP) should be offered during routine clinical practice but never forced upon the person. It is usually initiated in the out-patient or primary care setting following the diagnosis of a life limiting condition e.g. Huntington's disease, when there is a shift in treatment focus or before the person becomes acutely unwell (RCP 2009). It is best done at the initial stages of the disease while the person has the capacity to make decisions. However, the timing of discussions should be tailored to the readiness of the person and their individual needs giving respect to their personal values, unique cultural background and idiosyncratic beliefs.

ACP discussions need to be skilfully led and should be a process, not a single event or a tick box exercise (RCP 2009). This highlights the importance of effective communication which does not end with the documentation of the person's wishes but a continuous, ongoing discussion with them, the family and the multidisciplinary team.

Cultural and religious considerations

Discussions and decision making when planning for future care are rooted within cultural and religious beliefs, attitudes and values (Kataoka-Yahiro et al 2011). It may apply only to a particular culture and not shared by individuals and families from other cultures. The concept of autonomy, control, independence and individualism are dominant core values in Western societies (Volker 2005). Whereas Asian cultures consider family decisions as a function of the extended family and see it as a family event rather than an individual occurrence (Wilkinson, Wenger & Shugarman 2007). It can therefore be said that a person's culture explains the different receptivity to advance care planning. What is perceived as advantageous to one group may not hold the same beneficial value to another group with a different value system and different life experiences.


Identifying wishes and preferences


The foreseeable fatal impact of Huntington's disease provides a springboard for people to explore their options, identify their wishes and preferences and make arrangements for their future needs. This could include the following:

- **Wishes, preferences in relation to future treatment and care**
- **Feelings, beliefs, values that may influence preferences and decisions**
- **Preferred place of care and how this may affect the treatment options available e.g. home, hospital, hospice**
- **People that the person would like to be involved in decisions about care e.g. family members, significant others, legal proxies, Independent Mental Capacity Advocate (IMCA)**
- **Need for religious, spiritual, personal and practical support that helps maintain one's identity and dignity**
- **Interventions that may be undertaken or considered in an emergency e.g. CPR**
- **When appropriate, what the person wants to happen after they die e.g. handling of the body, organ donation**

Organ donation

Organ donation is when a person donates their organs for transplant. It would be helpful to explore the person's views about organ or tissue donation so that they can be facilitated to register and tell their family and friends that they want to be a donor. This would make it easier for them to agree to donation when the person is close to death.

The NHS Organ Donor Register is a confidential national database that holds the details of around 21 million people who want to donate their organs when they die. Further information about the donation process and about joining the NHS Organ Donation Register is available at the NHS Organ donation website  www.organdonation.nhs.uk.

Brain donation is of particular relevance in Huntington's disease research. At the Huntington's Disease Association, we can provide further information and advice - contact us on:  info@hda.org.uk

Formalised outcomes

Formalised outcomes of Advanced Care Planning (ACP) under the Mental Capacity Act (2005) include:

1. An Advance Decision to Refuse Treatment (ADRT)

ADRT is an option that can be made within the advance care planning discussion. It is made by the person who has the capacity to make decisions and / or with the help or support of a clinician. It only covers the refusal of a specified future life sustaining treatment. It must be written, signed and witnessed and should contain a statement that it applies even if the person's life is at risk. There is online training available to assist professionals to help people complete an ADRT – available at www.nhs.uk/conditions/end-of-life-care/advance-decision-to-refuse-treatment/

Specific issues that may need an ADRT include:

A. Clinically assisted nutrition and hydration

- Intravenous feeding
- Nasogastric tube (NGT) feeding
- Percutaneous endoscopic gastrostomy (PEG) feeding

People with Huntington's disease, their families and the MDT are often confronted with decisions around PEG tube insertion. The experience of choking badly and not receiving adequate nutrition and hydration usually make people change their minds about not having a PEG tube inserted. It is therefore important that the views of the patient and those close to them are listened to and considered (including any cultural and religious views). A clear explanation should be given regarding issues to be considered such as the benefits, burdens and risks so that they are enabled to make informed decisions in the early stages of the disease when they still have mental capacity and are able to communicate.

- Radiologically inserted gastrostomy (RIG) feeding
- Intravenous or subcutaneous infusion

B. Cardiopulmonary resuscitation (CPR)

CPR is an invasive intervention which include chest compressions, electric shock, injection of drugs and ventilation. Generally it has a very low success rate and its burdens and risks include rib fracture, damage to internal organs, hypoxic brain damage or increased physical disability. It is essential that the benefits, burdens and risks of this intervention should be considered in people with advanced Huntington's disease.

Any discussions and decisions made with the patient and those close to them should be documented in their record or advance care plan.

C. Artificial ventilation

D. Antibiotic treatment

2. The appointment of Lasting Power of Attorney (LPA)

A lasting power of attorney (LPA) is a legal document that lets the individual (the 'donor') appoint one or more people (known as 'attorneys') to help make decisions or make decisions on their behalf. This gives the person more control over what happens to them (as in the case of the diagnosis of Huntington's disease) when the time comes that they lack the mental capacity to make decisions.

Two types of LPA are:

- **For Health and Welfare**
- **For Property and Affairs**

For further information visit:  www.gov.uk/power-of-attorney

A property and affairs LPA may be used even if the person has capacity to make the decision.

Other advocacy services

1. Court Appointed Deputies (CADs)

They are individuals appointed by the Court of Protection where a series of decisions are required. They are helpful when a person's best interests require a deputy consulting with everyone and can make decisions on the person's behalf but cannot refuse or consent to life-sustaining treatments.

2. Independent Mental Capacity Advocates (IMCA)

They are part of a public consultation service for people with no other representative. They need only to be involved in specific decisions e.g. serious medical treatments; admissions to care homes or to hospitals. They can also advise on the person's best interests.

Involvement in research

Enroll-HD

Enroll-HD is an open-ended study that aims to collect clinical information about the person and their health. During each study visit they will undergo a series of movement and behavioural tests. Biological samples such as blood and DNA will also be collected. This may be more difficult during later stage Huntington's disease but other research opportunities exist.

Researchers will use this information and samples to learn more about Huntington's disease, to understand why and when certain symptoms appear and to try to find new treatments for the disease.

For more information visit  www.enroll-hd.org

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Symptoms



Symptom management

Effective management of symptoms in advanced Huntington's disease is crucial for people affected and their loved ones. However the disease complexity and duration of this phase of the condition can make this challenging. A person-centred, proactive approach which combines the expertise of all the relevant disciplines is necessary in order to meet the needs of people who have late stage Huntington's disease.

This section looks at symptoms commonly experienced by people with advanced Huntington's disease however the list is certainly not exhaustive. It is also important to always consider other factors if a person's symptoms suddenly change or deteriorate. For example, whether they could have an infection, are in physical pain or distressed, frustrated or anxious or whether there has been a change to their routine or environment.

Motor symptoms – chorea, stiffness, dystonia and rigidity

Motor symptoms in Huntington's disease include a mixture of both additional involuntary movements and increasingly impaired voluntary movements. The most common motor symptom in typical adult onset Huntington's disease is chorea but severity varies greatly from person to person. Early in the disease, people will generally appear more fidgety, restless and clumsy. In the late stages, chorea usually decreases and is largely replaced by bradykinesia (slow movements), stiffness, rigidity, dystonia (slow, abnormal movements and abnormal posturing with increased muscle tone) and sometimes myoclonus (sudden, often painful, shock like involuntary contractions of a muscle or group of muscles).

Some people with advanced disease do still have chorea however which can result in significant problems for them. The constant movements are exhausting and friction can result in skin breakdown. Safe moving and handling, particularly hoisting, can be very difficult when a person

has choreic movements and finding suitable seating, wheelchairs and beds can be challenging.

Stiffness, rigidity and dystonia can also cause significant pain and again can make washing, dressing and comfortable positioning very difficult. Jaw stiffness can further impede eating and drinking and can make mouth care hard to provide. People can additionally develop painful contractures.

There are a number of drugs that can be used to reduce chorea and other motor symptoms and these are summarised in the table on the next page. However most have side effects which may cause more problems than the symptom itself. It is essential therefore to assess the effect of symptoms such as chorea on the person's quality of life and to use caution when initially prescribing or increasing any of these drugs in people with advanced Huntington's disease. Additionally regular review of medication is vital so that they do not continue to take drugs which are no longer required or which may even be contributing to new symptoms such as increased stiffness.

Table 1: Commonly used medications for the management of motor symptoms (Novak & Tabrizi 2010)

Symptom	Medication	Notes and main side effects
Chorea	Tetrabenazine	Dopamine depleting agent. Can cause low mood, sedation
	Olanzapine	Atypical neuroleptic. Can cause sedation, parkinsonism, increased appetite and weight gain, raised blood glucose and lipids leading to increased risk of stroke in older patients. Risk of neuroleptic malignant syndrome. Helpful if patient also has irritability and agitation but can increase apathy
	Risperidone	As olanzapine, less effect on appetite
	Quetiapine	As olanzapine, less effect on lipids and glucose
	Sulpiride	Older neuroleptic. Can cause agitation, dystonia, sedation, akathasia, galactorrhea and amenorrhea
	Haloperidol	Older neuroleptic. Can cause sedation, parkinsonism, dystonia, akathasia, constipation, hypotension and risk of neuroleptic malignant syndrome
Myoclonus, chorea, dystonia, rigidity	Clonazepam	Benzodiazepine. May cause sedation, exacerbation of cognitive impairment, ataxia and further impair mobility
Rigidity	Levodopa	Amino acid precursor of dopamine. May cause gastrointestinal disturbance, postural hypotension, insomnia, agitation, psychiatric symptoms
Rigidity, spasticity	Baclofen Tizanidine	Skeletal muscle relaxants. May cause sedation, drowsiness, confusion, gastrointestinal disturbance, hypotension and further impair mobility

Botulinum Toxin (Botox) injections can also be very helpful in some patients who have muscle stiffness and spasticity and people can be referred to a specialist botox clinic for assessment.

Non pharmacological interventions play an equally important role in symptom management and are further discussed in the 'Quality of Life' section in this guide.

Table 2: Non pharmacological interventions for motor symptoms

Intervention	Useful for:
Physiotherapy	In advanced disease for example can provide advice on passive stretching and splinting to help prevent contractures.
Occupational therapy	Can help with safety and suitability of the home environment, advising on aids and equipment, suitable seating, padding, beds and lifting and handling equipment.
Wheelchair services	Can ensure that wheelchairs are suitable, for example are well padded to minimise injuries to patients with chorea and can recline or tilt to reduce the need for restraints such as seat belts. Also that they have adequate head support.

Behavioural problems

In the advanced stages of Huntington's disease, many of the difficult behaviours such as aggression, impulsivity and lack of insight become less problematic as the person becomes less physically able to act on impulse or out of frustration. However other behaviours such as resistive behaviour, agitation and screaming can occur and some people still experience episodes of aggression which can place themselves and/or others at risk.

As mentioned previously, if a person's behaviour changes suddenly it is always important to check for causes which may be exacerbating these symptoms such as infection or pain. If there are concerns that a person may have hit their head, it is very important that a head injury is ruled out.

Environmental factors which may be contributing to behavioural problems should always be considered and behaviour charts can be useful to identify any triggers or patterns. When a person is no longer able to communicate verbally, they can become frustrated and this can lead to agitation and challenging behaviour and so, where possible, finding out about that person – their likes, dislikes and preferences – can be helpful and alternative methods of communication can be considered. This is discussed in more detail in the section on 'Communication.'

Routine and a familiar, calm environment are often helpful to people

with advanced Huntington's disease and keeping to a routine and allowing enough time for activities such as personal care can help with resistive behaviour.

For agitation, benzodiazepines can be helpful however they should be used with caution due to potential side effects (see table 1). A review of medication can be useful – if a person tends to be more agitated at a certain time of the day, changing the timings of medications rather than increasing doses or adding in new medications can sometimes be enough.

Some people with advanced Huntington's disease will scream persistently and this can be extremely distressing particularly for caregivers and loved ones. When a person is unable to say why they are screaming, carers have to work through possible causes.

- **Is the person in pain, distressed, frustrated, depressed, hungry, thirsty or frightened?**
- **Is the room too dark or could they be experiencing hallucinations?**
- **Does the screaming stop when someone goes to the person and comforts them?**

Unfortunately it is often just not possible to identify an exact cause for the screaming, however strategies such as managing agitation, reviewing medications which may be causing side effects, treating possible pain and possible mood symptoms and providing reassurance and comfort to the person can be helpful.

Psychiatric symptoms

A wide range of psychiatric symptoms are seen in Huntington's disease including depression and anxiety, apathy, irritability and aggression, psychosis and obsessive/compulsive disorders. There is also an increased risk of suicide in people with Huntington's disease. Even in the advanced stage of the disease, people can experience psychiatric symptoms which have a profound effect on their quality of life and can exacerbate other symptoms. However assessment of mood is clearly much more difficult in someone with advanced Huntington's disease, particularly if the person is no longer able to communicate how they are feeling. Carers should be encouraged to be alert to changes in a person's facial expression, sleep pattern and appetite. Also if the person is crying out, has become more withdrawn or resistant to personal care

or leaving their room, these may be indicators of low mood. People who are still able to communicate may express feelings of hopelessness and helplessness and talk of suicide or people may report hearing voices or seeing things.

With any deterioration in mood, other factors should always be ruled out and the person's medications reviewed. The input of a Neuropsychiatrist who can work closely with and advise local mental health teams can be helpful in more complicated cases. People with Huntington's with depression should be treated in the same way as any other person with depression and Selective Serotonin Re-uptake Inhibitors (SSRIs) such as citalopram, sertraline, paroxetine and fluoxetine are often used. For people who are also experiencing insomnia with their depression, a more sedative antidepressant, such as mirtazapine, taken at night time can be helpful. Typical neuroleptics such as olanzapine and risperidone can be very helpful in managing irritability and aggression (see table 1). Likewise the other psychiatric symptoms including obsessive compulsive disorder, and psychosis should be treated according to recommended guidelines. Psychological therapy may be inappropriate for many people with advanced Huntington's disease however for some the opportunity to talk about their feelings, fears and frustrations can be helpful and can still be possible and so this option should not be automatically excluded just because of communication and/or cognitive impairment.

Pain

People with advanced Huntington's disease can experience pain for many reasons but as with other symptoms, it can be very difficult to assess when a person is unable to communicate and/or is cognitively impaired. 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 and palliative care specialists can advise further on such tools.

As mentioned previously spasticity, rigidity, stiffness and myoclonus can cause pain and should be treated accordingly (see table 1).

People with Huntington's can also experience neuropathic pain with or without any obvious structural cause and it may be advisable for the person to have appropriate imaging as advised by a neurologist or pain expert, although the necessity and potential benefits of this should be weighed up along with the risk of distress to the person. Effective

management with medications such as gabapentin, pregabalin and amitriptyline can often be achieved. In some cases treatment with opioid analgesics such as tramadol, oramorph and fentanyl patches may be necessary but these should be prescribed under the supervision of a palliative care or pain specialist. Again non-pharmacological interventions such as physiotherapy, postural management and complementary therapies can be useful.

It is important to remember that people with advanced Huntington's disease can experience pain for reasons not necessarily 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 so should be treated accordingly.

Elimination

In the advanced stages of Huntington's disease, people usually become incontinent of both urine and faeces and for their comfort and dignity, it is important that appropriate continence products are used. Regular pads may be ineffective, for example if a person has chorea. Some people require catheterisation and in those situations, a suprapubic catheter is often better.

People who are still continent often experience urinary frequency and urgency, which can necessitate frequent visits to the toilet. Antimuscarinic drugs such as oxybutynin and tolterodine can be helpful, however in men in particular other causes such as prostate problems should always be ruled out first and then potential side effects of medications carefully considered. A urology opinion can be helpful.

Urinary tract infections can regularly occur in people with advanced Huntington's disease and can lead to an acute exacerbation of Huntington's disease symptoms and so urgent treatment is extremely important. Keeping people adequately hydrated is important but can be difficult when they have impaired swallow and no PEG 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. Constipation should be managed 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 often profuse sweating and flushing. In the first instance, other potential causes for this such as infection, medication side effects or hormonal problems should be ruled out. Once other issues have been excluded, the most important thing is to try and manage this for the person as the feelings of heat can be very intense, uncomfortable and distressing and can lead to worsening of movement symptoms and agitation.

Some strategies include keeping surroundings – bedrooms for example – 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 or even bags of frozen peas! Clothing where possible should be light and made of natural fibres and t-shirts could be placed in a plastic bag in a freezer to cool them down before the person puts them on. Cold drinks and ice cream can be soothing for the person when they are feeling very hot or extra water can be given via a feeding tube and massaging levomenthol cream into the person's arms/legs can have a cooling effect.

In the very late stages of Huntington's disease, people can experience unexplained recurring high fevers where no infective cause is identified. Ensuring where possible that the person is hydrated is important and also giving the person simple paracetamol for example may help in reducing the fever as well as using the strategies above.

Hypersalivation

Many people with advanced Huntington's disease experience problems with excess salivation and drooling. This can be distressing for the them and cause difficulties with impaired swallow when 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. Non-pharmacological interventions include postural management and supports for the head and neck and manually assisted cough techniques.

Medications such as transdermal hyoscine (scopoderm patches), sublingual atropine 0.5% drops, glycopyrrolate, and ipratropium bromide inhalers can be helpful and palliative care specialists can further advise on these and appropriate doses. However people should be closely monitored to ensure that their mouths don't become too dry and additionally that other mucous membranes and skin don't become

dry. In cases where medication is not effective, botox injections may be helpful and a referral to a specialist botox clinic could be considered.

For people who have thick secretions, it is important where possible to ensure that they are well hydrated and humidifiers, steam inhalation and nebulisers can help.

Nausea and vomiting

Whether a person is still eating and drinking orally or alternative methods of feeding such as PEG feeding are being used, 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.

- **Is the person constipated or do they have an infection?**
- **Are they taking any medications which could be causing nausea as a side effect 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, it is important to review when the nausea and vomiting occurs. Does it occur directly after eating/drinking? If so, reviewing positioning and seating when the person is eating/drinking can be helpful. If the person is still eating orally, an up to date speech and language therapy review can be helpful. It may be that the person is eating too quickly due to hunger and impulsivity and therefore cramming food into their mouths and so caregivers need to remind them to eat slowly, taking one mouthful at a time. They may also be taking large gulps of fluids whilst eating which is 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, again a review of posture during feeds is important.**
- **If the person has overnight feeding via a pump, are they lying flat?**
- **Could the feed rate be too quick?**
- **Or is the feed suitable?**

A dietitian review can be very helpful as they can advise on adjusting feeding times, rates, volumes and feed types to try and minimise any feelings of nausea.

If the person is experiencing reflux, a proton pump inhibitor such as omeprazole or lansoprazole may be helpful. Antiemetic therapy can also be considered and medications such as levomepromazine or metoclopramide can be helpful.

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 although it is worth noting that myoclonic jerks can be mistaken for seizure activity. If there are any concerns that someone has had a first seizure then an urgent neurological opinion should be sought so that appropriate investigations can be performed and treatment instituted if necessary.

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Finances



Benefits

People with Huntington's disease may qualify for a range of benefits such as Personal Independence Payment (PIP) or Attendance Allowance (AA). If the person is expected to live less than six months because of Huntington's disease or another health condition then they can apply for the above under special rules (to ensure claims are dealt with more quickly). Those who are claiming under special rules should ask their GP to complete a DS1500 form.

Most benefits stop when people move into a care facility, the rules are different depending if it is a residential or nursing home and advice should be taken from the relevant advice lines.

There are a range of benefits which may be available to a spouse or civil partner of someone who has died (Bereavement payments, Widowed Parents Allowance, Bereavement Allowance) and advice may be sought from Citizens' Advice Bureau or local Welfare Rights team.

More information is available on all of the above from www.gov.uk

Bereavement Service helpline: ☎ 0345 606 0265 / 0345 606 0275
(Welsh language)

Care funding

The amount someone pays for long term care depends on their health, the level of support they need, their income and their savings and assets. A person may be required to pay for all of their care, some of it or nothing at all.

NHS Continuing Care: Those with a high level of nursing needs may be eligible for free Continuing Health Care which is arranged and funded solely by the NHS.

Local authority funding: The local authority may assist with the costs of residential care or help at home by providing support for carers, equipment and specialist services. Funding depends on:

- **Individual need (assessed via a care and support needs assessment)**
- **How much the person can afford (assessed via a financial assessment)**

In some circumstances, it is possible that funding for care and support is provided by the NHS and local authority (joint funding).

Self-funding: A person may not qualify for funding from the NHS or local authority. Or, the amount they receive may not cover the care costs in full.

More information is available from the Money Advice Service:

 www.moneyadviceservice.org.uk

Joint ownership/deferred payment

What happens if you're a home owner?

If a person owns their own home, it may be counted as capital 12 weeks after they move into a residential or nursing home on a permanent basis. There are circumstances where a home won't be counted as capital if certain people still live there (such as a spouse or civil partner).

Deferred Payment Agreements

After the first 12 weeks, any financial assistance the local authority provides can be regarded as a loan against the value of the former home and repaid from its eventual sale.

More information is available at  www.ageuk.org.uk

Losing a wage - a survivor finding a job

Finding a job

When caring for a dependant a family carer may have significant financial worries as they may have been surviving on benefits which have ceased following the death of their loved one. All state benefits that the person was receiving will end on that day, giving little time for them to grieve before having to find employment. There are many online employment agencies and Job Centre Plus is another source of help. The person may want to return to a previous occupation or career, or may wish to retrain.

Local carers' services should also be able to support them in making the transition back to work or training - some have special projects and support groups.

Funeral costs

A typical funeral using a funeral director costs around £4257 (Royal London National Funeral Cost Index Report 2017). However, it can be much more or less than this, depending how a person wants to remember the person who's died, what they can organise themselves and how much they can afford to pay.

People may be able to plan in advance for their own funeral by using an earmarked savings account or a funeral plan (though terms and costs should be checked carefully) and some insurance plans have a payout for funeral costs.

Funeral Payments may be available for those on a low income who need help to pay for a funeral they are arranging. These usually have to be paid back from the deceased person's estate if they have one.

British gas energy trust may help with funeral costs where outstanding funeral expenses are causing hardship.


More information is available from the Money Advice Service:

 www.moneyadviceservice.org.uk

Extra heating, washing, drying

At the advanced stages of Huntington's disease the person may spend most of the time in bed. This can lead to pressure area break down and there may be incontinence. Bed linen and bed clothes can be soiled which may mean daily washing and drying and the need to buy more sheets and nightwear. The person with Huntington's disease may have difficulty in controlling their body temperature which may either increase or decrease the need for heating the home.

It may also be possible to get cold weather and winter fuel payments. For further information visit  www.gov.uk

If a person's utilities supplier has signed up to the Safety Net for Vulnerable Customers Scheme they are committed to never knowingly disconnecting vulnerable customers. Where a customer has been disconnected and then is identified as vulnerable, the supplier will reconnect their customer as a priority. A person is considered to be vulnerable if for reasons of age, health, disability or severe financial insecurity, they are unable to safeguard their personal welfare or the personal welfare of other members of their household. More information is available at  www.energy-uk.org.uk/policy/disconnection.html

Prescriptions and products

NHS prescriptions

A person in England with Huntington's disease may get free NHS prescriptions as some people qualify under the following criteria 'have a continuing physical disability that prevents you from going out without help from another person and have a valid MedEx' (this is via a FP92A form which a doctor can sign). There may be other reasons why they qualify such as being over 60 or on a low income.

Non-prescription costs

The person with Huntington's disease may enjoy other therapies that are not covered by prescription such as aromatherapy, relaxation therapy, reiki and massage. These can be helpful but expensive, many hospices provide these free of charge. There may be useful pieces of equipment that are not provided by statutory services but grants may be available.

Incontinence products

The person may be able to get incontinence products on the NHS, such as incontinence pads or bedding, but it depends on the local NHS organisation. To qualify for incontinence products, they may need to:

- **be assessed and start a treatment plan**
- **meet criteria set out by the local clinical commissioning group (CCG)**

More information is available at www.nhs.uk

Hospital costs

There are occasions when travel to hospital can be arranged for appointments. Some Trusts have a voluntary service in place. If the person is an in-patient for a prolonged period of time, hospital car parking charges may be reduced or waived. This has been free in Wales since 2011 unless still under private contract.

In England, concessions, including free or reduced charges or caps, should be available for the following groups:

- **people with disabilities**
- **frequent outpatient attenders**
- **visitors with relatives who are gravely ill**
- **visitors to relatives who have an extended stay in hospital**

Details of charges, concessions and additional charges should be well publicised including at car park entrances, wherever payment is made and inside the hospital. They should also be included on the hospital website and on patient letters and forms, where appropriate.

Other charges such as bedside TV may be imposed when in hospital. This differs across the country.

When considering a care home a person should think about how family and friends can travel to visit the person with Huntington's disease. Travel costs can be expensive.

- **Is there a public transport route and is it regular?**
- **Does it cease at a certain time?**
- **What about evenings and weekends/bank holidays?**

More information is available at  www.gov.uk

Costs of future planning

Future planning is often especially important for someone with Huntington's disease and this is covered in other sections of this booklet. However, this can become very expensive.

There is a fee for registering a Lasting Power of Attorney, although the person may not have to pay the fees if they are on means-tested benefits or a low income.

Wills can also be expensive and there are different types but there are a number of ways a person can get a Will written. A solicitor is perhaps the most expensive, followed by a specialist Will writing service. The cheapest option is by buying a template document in a stationery shop and a person writing their own Will but they have to be careful about how they write it. A person's best option depends on how complicated their wishes are.

More information is available at  www.moneyadvice.service.org.uk

Quality of life



What is quality of life?

The Quality of Life research unit in Toronto describe quality of life as

“The degree to which a person enjoys the important possibilities of his/her life. Possibilities may result from the opportunities and limitations each person has in his/her life and reflects the interaction of personal and environmental factors.”

Quality of life can be defined into three categories which are:

- **Being - which encompasses physical, psychological, and spiritual needs.**
- **Belonging - which covers connections to the environment, social environment, access to resources and;**
- **Becoming - which involves purposeful activities, employment, volunteering and leisure activities.**

(Quality of Life research unit, University of Toronto)

It can be difficult when someone is in the later 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 the person does not understand what is happening around them. Research has shown that 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.

It is important to remember the need for continued stimulation. People with Huntington’s disease still have hobbies and interests and it is essential that they can pursue these. It is the care giver’s role to facilitate this. The involvement of an activity co-ordinator where possible can help assist with this. It is important to plan activities around their interests and to take into consideration their shorter concentration span and

the tendency to fatigue. The development and use of tools such as life story books, and gathering information from available family and friends about the person's hobbies and interests will help target activities that the person may want to engage with.

It is important to recognise that people with Huntington's disease will still have sexual needs and creating an environment that will enable them to facilitate this is essential. Carers also need to recognise that a person's intimate relationships may be affected by changes. In late stage Huntington's disease it may be difficult to establish what these needs are but it is important to think about them.

It is unlikely when someone is in the later stages of Huntington's disease that they will be able to have a proactive role in their care. Therefore any time spent on one to one activity that involves human touch should be seen as beneficial.

Looking after someone's personal hygiene in late 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 regular routine and talking through with the person each step of the way will be helpful. Oral hygiene and dental care are really important in later stage Huntington's disease. Ensuring that someone's mouth is empty following a meal, regular teeth cleaning with a specially adapted brush such as 'Dr Barman's Super Brush' as well as involving a community dentist for regular check ups, if visiting the surgery becomes difficult, are essential. Weight loss may also occur if ill-fitting dentures are worn, so these should be checked at regular intervals.

There are several reasons why people with late stage Huntington's disease are more susceptible to the development of pressure sores. These include weight loss, poor diet, either involuntary movements or rigidity and stiffness. It will be difficult to heal a pressure sore once formed so vigilance and regular pressure area care is essential. Referral to a physiotherapist and occupational therapist can help with the provision of appropriate equipment and positioning of the person. Chest physiotherapy in particular can be helpful in late stage Huntington's disease to reduce the risk of aspiration pneumonia.

Further Reading

There are some excellent guidelines for both produced by the European Huntington's Disease Network ('Guidelines for Occupational Therapy in HD,' www.futuremedicine.com/doi/full/10.2217/nmt.11.81)

'Physiotherapy clinical guidelines for Huntington's disease' and 'Development of physiotherapy guidance and treatment-based classifications for people with Huntington's disease,' www.hda.org.uk/media/1369/hd-physiotherapy-clinical-guidelines.pdf

There are many professionals involved in the care of someone with late stage Huntington's disease and sometimes this can generate lots of different hospital appointments which can prove to be a challenge. 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.

It can be difficult to really know and understand what a person was like before they had Huntington's disease, who they were, what was important to them, things that really mattered to them. It is essential to gather as much information as possible from family and friends if the first time the care giver meet the person is during the later stages of the illness, or from the person themselves earlier in the illness. This will help the caregiver for the person and help them to help them to maintain their identity. It can also be really useful to enable them to establish any particular significant people (or animals) that will form an important part of their lives. Establishing any last wishes or things that they wanted to do prior to death and helping the person achieve these can influence their quality of life. Having an awareness of someone's religious and spiritual needs and facilitating for these to be met can provide great 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. Family caregivers are often exhausted by the time a person goes into full time care. They may be left feeling guilty that they have not maintained that person at home. It is also essential to remember that often the family members are they themselves at risk of

developing the illness. Seeing their loved one progressing through the illness is a mirror image of what could happen to them. That presents a real challenge, ensuring the best possible care for both the person affected by Huntington's disease and their family.

People with late stage Huntington's disease can be particularly emotionally challenging. A family carer may have cared for them for many years and no longer be able to and professional carers may have also cared for and known the person over many years. Training and education about the disease, its symptoms, treatment and management is essential. Sometimes the progressive nature of the illness can lead to a feeling of helplessness. The death of the person can be very challenging. It is important family carers are offered support and should seek a carer's assessment, counselling and access bereavement support through organisations such as Cruse, or from the from our Specialist HD Advisers at the Huntington's Disease Association. For paid carers it is vital they are self-aware and seek the support from other colleagues and professionals. Being part of a team and using staff supervision can also help. The Specialist HD Adviser can also offer support to paid carers.

Perhaps the best way to sum up the importance of quality of life in Huntington's disease is by a poem written by Trish Dainton, carer for her husband with Huntington's disease and taken from a the book of poems she wrote, "Curse in Verse and Much More Worse."

The quality of life by Trish Dainton

This poem emphasises the need to consider the effect of illness on the whole family group. It's fair to say any illness one person has will have a knock on effect for those close. However, with its hereditary issues, and the complexity of the illness (physical, psychological and emotional), by treating peoples' needs in isolation the health service are likely to be wasting resources.

Back from the Doctors,
With Pills for HD.
Not just the patient,
But whole family!


There's dad with the gene,
And son who's depressed.
There's mum who can't sleep,
And daughter who's stressed.

The powers on high,
Do not seem to think,
Of savings they'd make,
If they just made the link!

The link ... The core focus,
Their Quality of Life;
The effective way forward,
For man, child and wife!

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Huntington's Disease Association website


 www.hda.org.uk/professionals/resources-for-professionals

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Get in touch

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Huntington's Disease Association

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