



# HUNTINGTON'S DISEASE ASSOCIATION



## STANDARDS of CARE

IN ASSOCIATION with



**St Andrew's**  
HEALTHCARE

# Huntington's Disease Association

## Standards of Care



# Foreword

The idea to develop some standards of care in Huntington's disease (HD) came about as a direct result of working with the many committed and enthusiastic professionals working in the field, seeking evidence to support their practice. It was always our intention to produce something that was practically based. We owe a debt of gratitude to those professionals who attended the focus group meetings and who provided us their ideas and expertise to work with.

I would like to acknowledge and thank the working group of regional care advisers that took this idea and concept and made it into a reality, through hard work and determination. A particular thanks to Ruth Sands, Helen Brewer, Mandy Ledbury and Diana King. Ruth in particular must be thanked for her determination to ensure that she received all of the information needed and put it into the current format.

I would also like to thank our sponsors who have made the production of this leaflet possible, a grateful thank you to St Andrew's Healthcare without whose help we would not have been able to produce this document

It has been an interesting project, and will be an ongoing project but it has been exciting and encouraging to see the dedication and knowledge of so many professionals working in the field of HD.

Cath Stanley. Head of Care Services. Huntingtons Disease Association.

# Acknowledgements

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There are many other people who have inputted their time and expertise to these Standards of Care, but it is too many to list here but they are not forgotten and we are extremely grateful for their input.

# Introduction – Standards of Care for people with HD.

## Developing the standards of care

This document is aimed at Social and Healthcare professionals; however anyone who is caring for someone with HD may find it useful. It has been put together by a small group of professionals with a special interest in HD.

The document aims to provide 'person centred' Standards of Care. 'Care' for people with HD is very much lacking in evidence based research; where possible these Standards are evidence based , where this hasn't been possible they are rely on the experience based practice of those with extensive experience of caring for people with HD.

Four main topics are covered in this first edition:

- Behaviour
- Risk
- Diet, Swallowing and Communication
- End of Life

These will be reviewed and updated and new topics added where it is deemed appropriate.

If you have any comments on these standards, or you would like to be involved in future updates / additions, please contact the HDA Head Office on T: 0151 298 3298 E: [info@hda.org.uk](mailto:info@hda.org.uk)

## *Disclaimer*

*These Standards of Care have been put together based on the knowledge and experience of clinicians who have worked with many patients with HD. These Standards can provide guidance but do not override the individual responsibility of healthcare professionals to use their clinical reasoning and make decisions appropriate to the individual they are working with.*

# About Huntington's Disease

Huntington's disease, which is often called HD, is an hereditary disorder of the central nervous system. It used to be known as Huntington's Chorea or HC. Huntington's disease usually develops in adulthood and can cause a very wide range of symptoms. It affects both men and women.

## What causes Huntington's disease?

Huntington's disease is caused by a faulty gene on chromosome 4. The gene, which produces a protein called Huntingtin, was discovered in 1993. In some way - which is not yet understood - the faulty gene leads to damage of the nerve cells in areas of the brain including the basal ganglia and cerebral cortex. This leads to gradual physical, mental and emotional changes.

Each person whose parent has Huntington's disease is born with a 50:50 chance of inheriting the faulty gene. Anyone who inherits the faulty gene will, at some stage, develop the disease. A genetic test is available from Regional Genetic Clinics throughout the country. This will usually be able to show whether someone has inherited the faulty gene, but it will not show the age at which they will develop the disease.

## The early symptoms of Huntington's disease

The symptoms of Huntington's disease usually develop when people are between 30-50 years old, although they can start much earlier or much later and can differ from person to person, even in the same family. Sometimes, the symptoms are present for a long time before a diagnosis of Huntington's disease is made. This is especially true when people are not aware that Huntington's disease is in their family.

The early symptoms include slight, uncontrollable muscular movements, stumbling and clumsiness, lack of concentration and short-term memory lapses, depression and changes of mood, sometimes including aggressive or anti-social behaviour.

Great strain is put on relationships if unexpected temper outbursts are directed towards the partner. The time before a diagnosis is made can be very confusing and frightening because people do not understand what is happening and why.

Some people who know they are at risk spend time searching for the first signs that they are developing the disease. They may worry about simple things like dropping a cup, forgetting a name or becoming unusually bad-tempered. Most people do these things occasionally - whether they are at risk from Huntington's disease or not

- so they could be worrying unnecessarily. Anyone who is concerned should have a word with their GP who may refer them to a neurologist for tests. These tests could include a number of simple assessments and possibly a brain scan. The genetic test referred to earlier may also be used to aid diagnosis.

## How Huntington's disease progresses

Later on in the illness people experience many different symptoms but these may include involuntary movements, difficulty in speech and swallowing, weight loss, emotional changes resulting in stubbornness, frustration, mood swings and depression. Cognitive changes that people experience result in a loss of drive,

initiative and organisational skills. This may result in the person appearing to be lazy. There may be difficulty in concentrating on more than one thing at a time.

Sometimes psychological problems, rather than the physical deterioration, cause more difficulties both for the person with Huntington's disease and their carer. Some changes are definitely part of the disease process although they may be made worse by other factors. It is depressing to have a serious illness and extremely frustrating not to be able to do things which previously seemed simple.

In the later stages of the disease full nursing care will be needed. Secondary illnesses, such as pneumonia, are often the actual cause of death.

### What treatment and help is available?

Currently there is no cure for the illness but there are many ways to manage symptoms effectively. Medication can be used to treat symptoms such as involuntary movements, depression and mood swings. Speech therapy can significantly improve speech and swallowing problems. A high calorie diet can prevent weight loss and improve symptoms such as involuntary movements or behavioural problems.

## The Huntington 's Disease Association

The Huntington's Disease Association exists to support people affected by Huntington's disease. The HDA also provides information and advice to professionals whose task it is to support Huntington's disease families.

### The Association has:

- A Central Information, Advice and Support Service.
- A Regional Care Advisory Service.
- Local Branches and Groups throughout the country.
- A Research grants Programme.

**Huntington's Disease Association, Neurosupport Centre, Norton Street, Liverpool, L3 8LR**

**T: 0151 298 3298    E: [info@hda.org.uk](mailto:info@hda.org.uk)    W: [www.hda.org.uk](http://www.hda.org.uk)**

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Huntington's Disease Association

Standards of Care

# Behaviour



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## Introduction

Behaviour is the external manifestation of the individual's cognition and emotion – observers see the behaviour and may make assumptions about the underlying emotion and cognition.

### What sort of behaviour are we likely to see in HD?

Common behaviours that we may see in HD include:

- Aggression to self, objects and other
- Verbally inappropriate behaviour
- Sexually inappropriate behaviour
- Impulsivity / compulsivity

N.B This is not an exhaustive list, many other behaviours may be exhibited. More information is given in a HDA Fact sheet on 'Behavioural Problems in HD' and 'Understanding Behaviour in Huntington's Disease' (see resources)

The behaviour of the person with HD may be an attempt to communicate something e.g. being in pain, frustration, anxiety, needs or preferences.

### What issues arise because of the behaviours we see in HD?

- People's motivation can be affected – the way they participate in activities and interact with other people
- People's self image can be affected
- In extreme cases behavioural issues can lead to an increased risk to self and others and, the person with HD may need to move into a supportive care setting
- Behaviour can impact on relationships with family, friends and carers, the person with HD may be perceived as lacking in empathy and being selfish
- Behaviour can affect the person's ability to drive

## Assessment of behaviour

- Basic behavioural assessments can be carried out by any member of team, different professionals will have different levels of skill

### Functional analysis Assessment (A behavioural assessment)

A common way to undertake an assessment is via a 'functional analysis assessment' whereby the carer will keep a systematic record of behaviour in a diary form using the following guidance:

- When it took place
- What occurred immediately before the behaviour (trigger or antecedent)
- What form the behaviour took
- What occurred immediately following the behaviour
- Any other factors that seemed relevant to the occurrence of that behaviour

This can prove extremely useful in treating or managing challenging behaviour as when these diaries are analysed over time they reveal common triggers or factors occurring after a behaviour that may be maintaining the behaviour. This can form the basis of a management plan by the carers avoiding or manipulating the triggers e.g. if the person always becomes aggressive when asked to take a shower – the carers should consider and negotiate with the person if

- what is being asked is now too demanding without assistance
- If they are in pain
- If they would prefer a bath
- They are uncomfortable with the way the carer is speaking to them

Factors occurring following behaviours can also be powerful in maintaining the behaviour and consideration should be given to these if relevant. E.g. someone making over-personal comments about another person's appearance ("she's really ugly") may be unaware at the disapproval of others or inappropriateness of this statement if it is simply ignored and may then continue to make such comments. A quiet word about how this statement made the other person feel may help the person consider their behaviour in future.

#### Overt Aggression Scale – modified for Neurorehabilitation

This tool which was developed at St. Andrews allows consideration of the frequency, severity, type, environmental precursors, antecedents and interventions associated with aggressive/risk behaviours, which can then be used to develop individualised programmes (see resources – journal articles).

#### The St Andrews Sexual Behaviour Assessment (SASBA)

As above but used for inappropriate sexual behaviours (see resources – journal articles).

#### Other types of assessment (areas which may affect behaviour)

- Assessment of mental health e.g. Hospital Anxiety and Depression Scale (HADS)
- Assessment of general cognition e.g. mini mental state, modified mini mental state, Addenbrooks cognitive assessment
- Specific tests of cognition e.g. verbal fluency

Assessment should include communication abilities and take into account any pre-existing problems e.g. eyesight or hearing, posture problems, pain, infections, hunger, constipation, medication and side effects. Also social factors e.g. expected visitors not arriving, adjustment to losses, substance misuse

#### When do you assess

- Assessment of behaviour before acceptance to a service is essential to enable a realistic decision about the referral
- Assessment needs to be ongoing, allowing behaviour to be tracked, consider:
  - Does the behaviour vary over time?
  - Does the behaviour respond and reduce when you initiate your management plan (if not you may need to go back to the drawing board and think again!)

#### When do you refer to more specialist services? Who would this referral go to?

- A clinical psychologist or psychiatrist has expertise in this area and may potentially be referred to in more difficult situations where the answer is not obvious
- People whose behaviour is causing a risk to self or others would be referred to specialist services e.g. Mental health services covering HD
- There are many Care homes in the UK which specialise in the care of people with HD; these can be very good at assessing and managing behaviour. There are also centres within the UK which treat frequent and extreme behaviour disorders
- If specialist services aren't available, then referral should be made to the adult mental health services

#### Management of behaviour

Consistency of management plan is important as well as ongoing assessment of effectiveness / outcome

#### Staff training

- Staff should receive training around the behavioural, emotional and cognitive consequences of HD. Often an explanation of the cognitive changes characteristic of HD helps the carer to understand the behaviour and facilitates management, if necessary, even if the behaviour itself

does not change. Enabling staff to recognise how the person with HD may be thinking, through this they can gain the most important skill in managing behaviour– empathy. Key concepts staff need to learn in training are:

- how to communicate with the person
  - structuring of the day
  - flexibility in response
- RAID is a relentlessly positive approach to dealing with extreme behaviours - training for this technique is available from the Association of Psychological Therapies (see resources)
  - Non-abusive psychological and Physical Intervention (NAPPI) - NAPPI is one of the UK's leading providers of aggression management training (see resources)
  - Please contact your local Regional Care Advisor for training from the Huntington's Disease. The HDA have two documents 'I have HD – This means' and 'Tips for making life easier' which can be useful for staff who are new to HD (see appendices)

#### Legislation in relation to Behaviour

- Mental health Act 1983. The Mental Health Act applies to the assessment, admission and treatment of patients with a mental disorder. Available to order from the Department of Health [http://www.dh.gov.uk/en/Publicationsandstatistics/Legislation/Actsandbills/DH\\_4002034](http://www.dh.gov.uk/en/Publicationsandstatistics/Legislation/Actsandbills/DH_4002034)
- *Mental Capacity Act 2005*. The Mental Capacity Act (along with the Code of Conduct) provide both the statute and framework for acting and making decisions on behalf of individuals who lack mental capacity. Department of Constitutional Affairs [http://www.opsi.gov.uk/acts/acts2005/pdf/ukpga\\_20050009\\_en.pdf](http://www.opsi.gov.uk/acts/acts2005/pdf/ukpga_20050009_en.pdf)

## Resources Available in relation to behaviour

### HDA Materials

HDA Fact Sheet No. 10 Behavioural Problems

Rosenblatt, A., Ranen, N.G., Nance, M.A. and Paulsen, J.S. (1999) *A Physician's Guide to the Management of Huntington's Disease. Pharmacological and Non-Pharmacological Interventions*. Huntington's Disease Association of America. (Sections entitled the cognitive disorder / the psychiatric provide information about pharmacological and non-pharmacological management)

Understanding Behaviour in Huntington's Disease, Huntington's Disease Association

### Books

Quarrell, O. (1999) *Huntington's Disease The Facts*. Oxford University Press, Oxford. Chapter 3 'Behavioural and Emotional Aspects of Huntington's Disease'.

### Other Guidelines

Scottish Huntington's Association Draft Care Pathway (describes a managed care network approach and has sections describing cognitive/behavioural/mental health symptoms and their management).

### Journal Articles

Alderman, N., Knight, C. And Morgan, C. (1997). *Use of a modified version of the Overt Aggression Scale in the measurement and assessment of aggressive behaviours following brain injury*. *Brain Injury*. 11 (7) 503-523

Kingma, E.M., Duijan.E.V., Timman, R.,Van der Mast, R.C. and Roos, R.A.C (2008) *Behavioural Problems in Huntington's Disease Using the Problem Behaviours Assessment*. *General Hospital Psychiatry* (30) 155-161

Knight, C., Alderman, N., Johnson, C., Green , S., Birkett-Swan, L. And Yorston, G. (2008). *The St Andrews Sexual Behaviour Assessment (SASBA): Development of a standardised recording instrument for the measurement and assessment of challenging sexual behaviour in people with progressive and acquired neurological impairment*. *Neuropsychological Rehabilitation*. 18 (2) 129-159

### Guidance

The following Mental Capacity Act Booklets are available from The Office of the Public Guardian:  
<http://www.publicguardian.gov.uk/mca/additional-publicationsa-newsletters.htm>

- About your health, welfare or finance - who decides when you can't?
- A guide for family, friends and other unpaid carers

- A guide for people who work in health and social care
- A guide for advice workers
- The Mental Capacity Act - Easyread
- The Independent Mental Capacity Advocate (IMCA) service

#### Internet Resources

- **Non-abusive psychological and Physical Intervention (NAPPI)** - NAPPI is one of the UK's leading providers of aggression management training. <http://www.nappiuk.com/index.php>
- **The Association for Psychological Therapies (APT):** The APT provides training in mental health and related fields. Delegates on courses typically comprise: psychiatric nurses, social workers, occupational therapists, clinical psychologists, psychiatrists and others. <http://www.apr.ac/>

# Appendices

Appendix 1\_ I have HD – this means

Appendix 2\_Tips for making life easier



## Huntington's Disease Association

### I have HD – this means:

- I like my own routine
- I do **one** thing at a time
- You need to get my attention and then tell me what you want
- Give me time to answer – don't repeat what you said or put it another way – this makes it difficult for me to answer
- Listen to what I say – it takes a lot of effort
- I don't know how to **wait!** If I need something I need it **now**
- I need lots of little snacks and drinks
- My brain gets stuck on thinking about important things – so I repeat the same words a lot
- There is only **one** solution to a problem / question
- I remember my life before I was like this
- Sometimes I'm scared of the future – I think a lot
- I can still enjoy things and have fun
- I used to be independent and have a 'normal' life and make my own decisions – I don't want to change more than I have to



## Huntington's Disease Association

### Tips for making life easier.....

- Provide a calm predictable environment – establish a routine for the day that the individual is comfortable with
- Are there any 'trigger' factors to the person's mood / behaviour – what makes it worse / better?
- Keep it simple – don't overload the person with information / stimulation
- One thing at a time
- Problems with 'getting started' or initiation – use prompts – verbal / environmental
- Problems with 'getting stuck' or perseverance – consistent staff approach, establish boundaries, use the need for routine, provide alternatives
- Decrease the complexity of the individuals environment – keep things in obvious places – break things down into manageable tasks
- Encourage joint participation in activities
- Use short sentences, cues, pictures to communicate
- Allow increased time for the individual to respond to you – repeat and rephrase if needed
- Use external memory aids – whiteboards, calendars, clocks
- Remember there are symptoms of HD we cannot change – we have to be flexible, adaptable and creative in the care we give

# Huntington's Disease Association

## Standards of Care

# Risk



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## Introduction

Risk can be seen as the possibility of harm to others or oneself which can be raised by chosen actions

### Potential areas of risk in HD

- Food/ Nutritional – Choking, malnourishment, outdated food, aspiration
- Physical / posture – falls, pressure sores
- Fire risks – dropping cigarettes, leaving the gas on to light cigarettes
- Impulsive behaviour
- Protection Of Vulnerable Adults issues / Child Protection Issues
- Aggression to others
- Inappropriate sexual behaviour
- Dangerous driving
- Risk of using public transport

N.B this is not an exhaustive list, but these are some commonly seen areas of risk in people with HD.

### Who is it a risk to? Who is at risk?

We all have risk in our lives and choose to put ourselves at different levels of risk e.g. sky diving. We use our knowledge and understanding to make an informed choice about the risk in our lives.

People with HD may not always recognise that they are putting themselves in a risky situation; they might not understand the outcomes / consequences of their actions.

### What is an acceptable risk?

The level of risk may be perceived to be unacceptable where it may constitute harm to oneself or to others.

## Assessment of Risk

- An assessment of risk should consider the consequences of both engaging in the risk taking behaviour or not
- A formal risk assessment could be considered such as The DICES Risk Assessment and Management System, from the Association for Psychological Therapies (see references)

### Who should assess risk?

Different professions may be involved in the assessment of risk e.g.

- A social worker to assess environmental risks, both internally and externally. Threadbare carpets, heat, light, personal care/neglect, vulnerability, finances, stair/step hazards, animals (pets and vermin), physical and mental health, ability and disability etc
- A Speech and Language therapist to assess swallow risk
- A Dietitian to assess risk of malnutrition
- An Occupational Therapist / Physiotherapist to assess the risk of falling
- An occupational therapist to assess a person using public transport
- A nurse to assess skin viability / pressure sore risk
- The fire service for an assessment of fire safety within the home
- When assessing capacity a 'decision maker' should be identified, this would be a professional who has the confidence to assess. A neuro-psychiatrist or neuro-psychologist will have special skills in dealing with people with cognitive impairment and addressing issues of capacity such as capacity assessments. When considering issues of capacity there are useful guidelines to refer to namely the 'code of practice' and mental capacity act booklets (see references)

### Management of Risk

The aforementioned professionals will be involved in the management of that area of risk

- The level of risk for the person with HD needs to be regularly monitored, and the views of the person with HD taken on board. They may assess their level of risk very differently to professionals / family
- It is often assessed that there are many areas which need to be changed to minimise risk, these need to be dealt with sensitively. The issues should be prioritised so the person with HD doesn't have to cope with too many changes at the same time. When focusing on a solution a more positive outcome is more likely to occur when the person with HD has been given time to consider and adjust to the changes
- Boundaries can be discussed with the person as to what is acceptable, what isn't and what the consequences will be if their risk levels are unacceptable
- The aim is to avoid the need for crisis management by supporting the person with regular reviews

- If the person is in the community decisions have to be made as to whether they should be in residential care. Ideally the person themselves would make this decision by having an input into where they would go, visiting homes, spending time in respite before committing to a long term move

#### Equipment and resources to manage risk

- A good care package can reduce risks dramatically for the person with HD. Ideally care packages are flexible so the person can be assisted in various activities in and out of the home that have risks attached to them
- Driving – The Forum of Mobility Centres is a network of independent organisations who offer professional, high quality information, advice and assessment to individuals who have a medical condition which may affect their ability to drive, access or egress a motor vehicle (see resources)
- Minimise risk where possible with equipment e.g.
  - Smoking – A smoking aid is available for people with involuntary movements meaning that they can't successfully hold a cigarette. These aids consist of a weighted ash tray, into which the cigarette is fixed with a flexible tube and mouthpiece connected. The cigarette burns safely down to the holder. Also available – fireproof aprons, fire alarms, heat detectors
  - Specialist bed sides – specialist bed sides are available for people with severe involuntary movements who are likely to get limbs trapped if using 'standard cot sides'
  - Walking aids – The Occupational Therapist / Physiotherapist can advise on walking aids, some people with HD like to use a walking stick early in the disease process to highlight the fact that they have a disability. Some people manage well with rolator frames or gutter frames. N.B many people with HD experience difficulty with walking aids due to poor co-ordination and inability to multi-task. In some cases walking aids can increase a person's risk of falling
  - Padding – Elbow and knee padding can be useful for people who are continually banging extremities
  - Personal alarms – Useful where someone is at risk of falling; the individual can push a button / pull a cord in the home to get through to a response centre for assistance
  - Identification cards – these are useful for a person to show people that they have HD and this could be the reason for unusual behaviour / movements. These are available from the HDA Head Office (see appendices)

- Hoists – Mobile hoists often aren't appropriate for people with HD as there are a lot of parts to hit against if the person has involuntary movements, if a hoist is necessary, tracking hoists are normally the preferred option. People can also struggle with a standard sling as they don't provide enough support
- This is not an exhaustive list there are many other pieces of equipment to decrease risk in activities of daily living. The Disabled Living Foundation are a useful resource when looking for specialist equipment (resources)
- Funding for a lot of this equipment should come through Social Services, otherwise grants from various agencies may be required – the HDA have a small grant fund which could be used to purchase equipment

#### Staff Training

- Without a proper understanding of HD carers may in fact unwittingly increase the risk when trying to reduce it. Issues of risk need to be discussed as a whole team and decisions recorded in the care plan
- It is essential that carers are given training on HD and are given chance to discuss their concerns about risk in a supportive environment. Please contact your local HDA Regional Care Advisor for training sessions

#### Legislation in relation to risk

- Mental health Act 1983. The Mental Health Act applies to the assessment, admission and treatment of patients with a mental disorder. Available to order from the Department of Health [http://www.dh.gov.uk/en/Publicationsandstatistics/Legislation/Actsandbills/DH\\_4002034](http://www.dh.gov.uk/en/Publicationsandstatistics/Legislation/Actsandbills/DH_4002034)
- *Mental Capacity Act 2005*. The Mental Capacity Act (along with the Code of Conduct) provide both the statute and framework for acting and making decisions on behalf of individuals who lack mental capacity. Department of Constitutional Affairs [http://www.opsi.gov.uk/acts/acts2005/pdf/ukpga\\_20050009\\_en.pdf](http://www.opsi.gov.uk/acts/acts2005/pdf/ukpga_20050009_en.pdf)

## Resources available in the area of risk

### HDA Materials

HDA Fact Sheet No. 13 Huntington's Disease and Driving

HDA Fact Sheet No 15. Seating, Equipment and Adaptations

### Journals

Johnson, C., Knight, C and Alderman, N (2006) Challenges associated with the definition and assessment of inappropriate sexual behaviour amongst individuals with an acquired neurological impairment. *Brain injury* 20(7), 687-693

### Guidance

- The following Mental Capacity Act Booklets are available from The Office of the Public Guardian:  
<http://www.publicguardian.gov.uk/mca/additional-publicationsa-newsletters.htm>
  - About your health, welfare or finance - who decides when you can't?
  - A guide for family, friends and other unpaid carers
  - A guide for people who work in health and social care
  - A guide for advice workers
  - The Mental Capacity Act - Easyread
  - The Independent Mental Capacity Advocate (IMCA) service
  
- Department of Constitutional Affairs. *Mental Capacity Act 2005*. Code of Practice.  
[http://www.opsi.gov.uk/acts/acts2005/related/ukpgacop\\_20050009\\_en.pdf](http://www.opsi.gov.uk/acts/acts2005/related/ukpgacop_20050009_en.pdf)

### Internet Resources

- **The Disabled Living Foundation (DLF):** The DLF is a national charity that provides free, impartial advice about all types of disability equipment and mobility products for older and disabled people, their carers and families. W: [www.dlf.org.uk/](http://www.dlf.org.uk/) T: 0845 1309177

- **The Association for Psychological Therapies (APT):** The APT provide training in mental health and related fields. Delegates on courses typically comprise: psychiatric nurses, social workers, occupational therapists, clinical psychologists, psychiatrists and others. <http://www.appt.ac/>
- **The Forum of Mobility Centres:** A network of 17 independent organisations covering England, Scotland, Wales and Northern Ireland, who offer professional, high quality information, advice and assessment to individuals who have a medical condition or are recovering from an accident or injury which may affect their ability to drive, access or egress a motor vehicle. <http://www.mobility-centres.org.uk/>
- **The Driver and Licensing Agency (DVLA):** Provides medical rules, advisory information and guidance for drivers. <http://www.dvla.gov.uk/>

# Appendices

Appendix 1 \_HDA ID card

My Name is:

I have Huntington's Disease – a progressive condition of the central nervous system. I may be slow or unsteady on my feet, I may have difficulty speaking and writing clearly, but I can hear and understand please allow me time to manoeuvre and communicate.

**In case of emergency please contact:**

My Next of Kin:

Tel:

Doctor:

Tel:

Huntington's Disease Association 0151 298 3298

Registered Charity No. 296453

Huntington's Disease Association

Standards of Care

# Diet, Swallowing and Communication



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## Nutrition and Swallowing - Introduction

### Why is food, nutrition and swallowing important?

Meals should be social occasions when we spend quality time with one another and enjoy what we eat and drink! For carers they offer an opportunity to take the weight of your feet and spend quality time with your client / patient / service user.

Any of us could choke or aspirate (take food / liquids into the lungs) when eating / drinking but HD increases these risks. Speech and Language therapists are concerned with managing the risk so that the person with HD can eat / drink safely, effectively and with enjoyment.

It is important to encourage people to have a well balanced diet which includes all the essential nutrients, and where this is not possible, e.g. because of personal choice, to advise on suitable supplements

- To prevent people becoming underweight and / or regain lost weight
- To maintain social interaction

### Maintaining weight and calorie requirements in HD

As with everything else to do with HD calorie requirements vary from person to person, even within a family. Everyone who is involved with HD is aware that there are increased calorie requirements, unfortunately, in these days of evidence based practice, very few studies have been carried out to prove it. Those that have been done, do demonstrate that there are increased requirements and that the use of supplements can be beneficial, but the number of studies carried out are very small and did not involve many subjects (see references).

While calorie requirements are increased, the majority of people with HD do not require 5,000-6,000 calories per day, as used to be suggested. Some do have very high requirements, others however maintain their weight on a moderately increased calorie intake, while a few are overweight on a "normal" intake. Each individual's requirements appear to vary throughout the course of their disease process, therefore regular review and monitoring is essential as "prevention is better than cure".

Ideally people's weight should be maintained at the top end of the healthy range i.e. BMI 24 -26, if the person's weight is above this, implications on their health need to be considered. However if people are losing weight or are underweight (BMI less than 20) it is essential to promote weight gain and/or prevent further weight loss.

This is because individuals who are underweight are more likely to :

- Lose muscle mass and therefore feel weaker

- Catch infections and take longer to recover
- Develop pressure ulcers if their mobility is reduced
- Take longer to recover from illness, operations or wounds
- Become apathetic and depressed

There is some anecdotal evidence which shows that providing a high calorie intake can help to:

- Reduce chorea movements
- Improve cognition
- Improve speech & swallowing

Improvements are mainly seen in patients who are undernourished

### Assessment of Nutrition

Ideally there should be early referral to a Dietitian who can:

- Explain why diet is important and start to build a relationship with the individual and their carer.
- Find out what food the individual likes
- Assess current dietary intake e.g. by taking a diet history or asking for 3-4 day food diary – see appendix 1 (N.B. it is not always possible to make an accurate assessment as people are often vague about what they eat when seen in clinic or at home, and completing a food diary often depends on the person or their partner/carer being willing and able to do so)
- Discuss their current dietary intake in a positive way and negotiate making changes if necessary. Dietary advice will need to be tailored to each individual, but these are some overall points to take into consideration
  - Aim to improve overall balance
  - Increase calories if necessary, using “food first” suggestions initially (see below) and using supplements when required
  - If diet is poor it may be necessary to negotiate small gradual changes that the person is willing and able to make, and to accept that their diet may never be ideal. This could be because of personal choices or because of physical or other limitations due to their HD. It is important to work with individuals and their carers to encourage appropriate changes if necessary and the use of suitable supplements if they are required.

- Remember to take into account their partner/carer's dietary requirements e.g. do they have diabetes, raised cholesterol levels, and their abilities
- Check weight, height and weight history
- Arrange for appropriate review and give contact details
- Regular follow up is important, the frequency will vary according to each patient, individuals in the early stages may only need to be reviewed once a year, but as the disease progresses this will need to be increased. Those who are more affected or whose weight is giving cause for concern may need to be reviewed as often as monthly

### Management of Nutrition

Maintaining good nutrition involves multidisciplinary team work and should involve

- Speech and Language Therapists
- Dietitian
- Occupational Therapist
- Physiotherapist
- carers
- catering staff
- MOST important the person themselves

### Anticipate problems and take early action

- Keep an eye on the individual's weight; if weight loss occurs (or weight does not increase) refer to a Dietician if they are not already seeing one
- Prompt individuals to attend meal times
- If a person feeds themselves and a lot of food is being wasted due to spillage, consider increasing the portion size to compensate
- Firstly try adding extra calories. "Food First"
  - Food is a good source of nutrients – remember people may need to eat differently if they are under nourished, i.e. "healthy eating" messages may not be appropriate & requirements for some nutrients are increased

- Eating is 'normal' rather than medical
- Ordinary food is usually more enjoyable than taking sip feeds etc.
- Eating may have social and psychological benefits

### Ways of adding extra calories

Snacks between meals e.g:

- Cheese and biscuits
- Digestive biscuits with butter
- Sandwiches with fillings such as: cream cheese, peanut butter, jam, mashed banana and evaporated milk
- Milky drinks e.g. milk shakes, smoothies, drinking chocolate
- Sweet biscuits & cakes
- Crumpets & butter
- Thick and creamy yoghurts, mousses, fromage frais
- Use plenty of butter, jam etc on bread, toast, crumpets etc
- Add plenty of butter, cheese, evaporated milk, salad cream to potatoes and vegetables
- Encourage high calorie puddings and add extra cream, evaporated milk, ice cream etc
- Use fortified milk for drinks and cereals (4 tablespoons milk powder to 1 pint full cream milk)

If "food first" does not work or is not appropriate then prescribable supplements should be used, these come in a variety of forms, and are available on prescription according to NICE guidelines because of disease related malnutrition

- Nutritionally complete sip feeds, either milk, yoghurt style or juice varieties
- Supplement puddings
- Milk shake powders
- High Calorie powders
- Protein and calorie powders
- High Calorie Liquids

Even if a Dietitian has not been involved in someone's care in the early stages they should be involved at this stage to decide on which supplements would be the most appropriate for each individual person.

### Nutrition management – PEG feeding

#### When should PEG /Advance Decisions to Refuse Treatment (ADRT) be considered?

When to discuss PEG feeding will vary from person to person, and will depend on many factors e.g.

- Their stage in the disease
- Personal experiences
- How they are coping with their diagnosis
- Their current physical condition
- Their carer's feelings

Ideally it should be introduced fairly early on if possible, and discussed in more detail when the individual and their carer feel they want more information. It should be discussed by someone who has built up a good rapport with the individual and their carer, however they need to be able to answer questions accurately, honestly and without allowing personal preferences to be apparent. There is possibly a need for some form of training to give people the skills and confidence to be aware of when and how to discuss PEG feeding.

If an individual decides to put an ADRT in place it needs to be shown that they have the capacity to make that decision and they need to be aware that if in the future they wish to change their mind they can do so. Once an ADRT is in place it is essential that all those involved in that person's care are informed and that they support that decision, regardless of their own opinions. For more information see the End of Life standard.

If an individual's swallow becomes unsafe but there is no ADRT in place and they are judged to be unable to make a decision, the placing of a PEG should be discussed with their next of kin/carer as they may be aware of any long standing opinions that the individual may have held. Health professionals need to avoid putting pressure on the next of kin/carer to persuade them to agree to something that the individual would not have wanted, if they are certain they had definite opinions either way.

If an individual has an ADRT in place then there also needs to be an end of life care pathway so that everyone involved in their care knows what to do in the event of them choking or developing a chest infection or aspiration pneumonia. This is essential to prevent admission to hospital for active treatment if it is against an individual's wishes.

### What happens if someone changes their mind / pulls out the PEG

If an individual changes their mind about not wishing to have a PEG this should not be a problem, the difficulty comes if they have one and wish to have it removed or they decide they do not wish to have one when it becomes necessary. It will need to be ascertained whether or not they have the capacity to make that decision.

If an individual is continually pulling their PEG out and it in distress when it is replaced this would seem to indicate that they no longer wish to have it. However there can be any hard and fast rule and that each individual case would be to be looked at separately, This is can be grey area if someone is unable to communicate and would need to involve the whole team and their next of kin/carer to reach a decision. It may be possible to delay re-siting the PEG to see how the individual manages?

### Assessment of Swallowing

As soon as speech becomes impaired (dysarthric) it is a good idea to have a Speech and language assessment of the oral management of food, since the tongue co-ordination is essential to both speaking and eating / drinking.

### Management of swallowing

- Feeding someone with HD should be regarded as a highly skilled activity, requiring information and support from the Multi Disciplinary team about positioning, amount, texture (food and fluids) utensils and timing for each person to be fed
- Self feeding is usually safer than being fed. People can be enabled to self-feed by careful positioning, the use of adapted cutlery, cups and dishes and/or hand-over-hand support
- The amount of food / fluids taken by someone with HD (advised by the dietician) may be affected by the texture of their meals / drinks – to be advised by the Speech and Language therapist
- Puree is not necessarily the most manageable texture for someone with HD. Generally a moist-mash (i.e. smooth and moist texture without lumps) is easier to eat, unless the Speech and Language Therapist Advises otherwise
- Speech and Language Therapists advice on thickening drinks and medicines should be strictly adhered to by carers, because of the risk of aspiration of fluids
- Many people with HD find ice-cold fluids easier to manage than those at room-temperature, as they give more sensory input to help control the fluid. (“Ice-cold” here means as cold as a Slush Puppi!)

- Fatigue becomes an increasing issue in mealtime management as HD progresses. Muscles affected by HD become tired quickly, especially when engaged in repeated, fast movements like those for speech or eating/drinking
- At this stage, when fatigue makes eating/drinking more risky, it is usually better to have several smaller meals each day, rather than 3 large meals
- Meals should not continue for more than 20 minutes of continuous feeding – a break of 10 minutes between courses may help
- Medication for their HD could cause **reflux**- when some stomach contents come back up the gullet/oesophagus. Reflux can be aspirated so people with HD should be encouraged to sit upright for at least 20 minutes after eating/drinking

### Communication – Introduction

#### What Problems is someone with HD likely to have with their communication?

People with HD are likely to show impaired language processing and along with dysarthria:

#### Impaired Language Processing

- Comprehension of words is generally retained but slows down, this affects the memory of the message so “information overload” happens more easily
- Retrieval of vocabulary is impaired (word finding difficulty), this impacts on speed and accuracy of responses / information – giving
- Verbal communication generally becomes more effortful

#### Dysarthria

- Chorea and impaired muscle tone result in:
  - Faulty co-ordination of breathing and voice, affecting volume and pacing of speech
  - Co-ordination of larynx impaired, affecting vocal pitch
  - Reduced co-ordination of oral and facial muscles, affecting speech sounds and facial expressions
  - Unpredictable speech patterns, further reduce intelligibility

A person's level of communication is likely to vary dependent on:

- The environment
- If they are tired
- Their current mood
- Their current health
- The reaction of other people

Communication difficulties can add to the social isolation that people with HD often experience.

#### Assessment of communication

An assessment needs to be carried out to establish the individual's skills in verbal communication and assess whether other means of communication need to be put in place. The assessment should be carried out by a Speech and Language Therapist and referral to this team should be early in the disease process.

#### Management of communication

Communication systems must be simple and appropriate

- Low tech approaches
  - Activity charts / weekly planners
  - Communication boards / charts
  - Life books
  - Talking mats

Strategies for communication:

- Speak slower and make the message simple
- Ask the person to repeat themselves
- Use yes/no questions
- Reduce distractions
- Use non-verbal communication when appropriate

- Communication becomes particularly difficult in the later stages of the disease when the person's verbal communication can be minimal. People with HD normally maintain understanding throughout the disease even when they can't make their wishes known. When communication is minimal it can be useful to:
  - Remember what the person had previously asked for and continue to provide these
  - Aim to establish a routine so the person knows what to expect
  - Explain what you are doing

N.B This is not an exhaustive list. More information is given in a HDA Fact sheet on 'Communication Skills' (see resources)

## Literature in relation to Diet Communication and Swallowing

### HDA Materials

HDA Fact Sheet No. 6 Eating and Swallowing Difficulties

HDA Fact Sheet No. 7 Huntington's Disease and Diet

HDA Fact Sheet No. 9 Communication Skills

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# Appendices

Appendix 1\_Nutrition Assessment Tool with kind thanks to Nottingham Community Nutrition and Dietetic Service

Appendix 2\_Basic Information about Gastrostomy (PEG) Tubes and Feeding. With kind thanks to Nottingham Community Nutrition and Dietetic Service

Nutrition Assessment Tool				
Name	I.D. Number	Date	Time	
Please complete all questions				Tick if action required
1 Do you eat?				
Breakfast	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	<input type="checkbox"/>
Lunch/midday meal	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	
Tea/evening meal	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	
Snacks between meals	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	
2. Do you have a cooked meal daily ?				
	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	<input type="checkbox"/>
Comments (e.g. who cooks the meals)				
.....				
.....				
3.Do you have a drink?				
With every meal	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	<input type="checkbox"/>
Between meals	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	<input type="checkbox"/>
During the evening	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Sometimes <input type="checkbox"/>	
4. What do you drink and how much?				
.....				<input type="checkbox"/>
.....				
5. Have you unintentionally lost or gained weight recently ?				
Gained weight	Yes <input type="checkbox"/>	No <input type="checkbox"/>		<input type="checkbox"/>
Lost weight	Yes <input type="checkbox"/>	No <input type="checkbox"/>		<input type="checkbox"/>
Comments.....				
.....				
6. Do you have any difficulties eating?				
	Yes <input type="checkbox"/>	No <input type="checkbox"/>		<input type="checkbox"/>
Why?.....				
7. Are you on a special diet because of a medical problem				
	Yes <input type="checkbox"/>	No <input type="checkbox"/>		<input type="checkbox"/>
Why?.....				
8. Are you taking any food, drink or vitamin supplements? (e.g. Complan, Build Up Fortisip)				
	Yes <input type="checkbox"/>	No <input type="checkbox"/>		<input type="checkbox"/>
<u>Please make a note of any actions taken on the reverse</u>				

Basic information about Gastrostomy (PEG) Tubes and feeding

**Introduction**

This leaflet has been put together to give you some basic information about gastrostomy tubes. It explains how the tube works, and answers some of the questions that are often asked when discussing gastrostomy feeding.

If you ever need to have a gastrostomy you will be given more detailed information at the time.

**A summary of important points.**

- ◆ A gastrostomy tube goes straight into the stomach
- ◆ A gastrostomy tube is used to give a liquid feed that will provide enough nutrition and fluids to maintain someone's health.
- ◆ A tube does not stop you eating and drinking if you are able to do so.
- ◆ Life will probably be longer with a tube in place.
- ◆ A tube will not cure Huntington's Disease or prevent the disease progressing
- ◆ A general anaesthetic is not needed to have the gastrostomy tube placed
- ◆ The tube is discreet and tucks under clothing
- ◆ Nothing other than liquid feed, water or liquid medication should be put down the tube
- ◆ Always inform the dietitian if any problems arise with the tube or feeding regime

**Why is a gastrostomy usually called a 'PEG'?**

The initials PEG stand for Percutaneous Endoscopic Gastrostomy, which describes the way the feeding tube is inserted into the stomach through the skin using an endoscope. An endoscope is a flexible instrument used to look inside people's stomachs by passing it through the mouth, down the throat and into the stomach. The feeding tube (PEG) allows you to receive the nourishment you need without having to take food or fluids by mouth, although it may still be possible to have small amounts of food and drink by mouth if you wish.

**Why may I need a PEG tube?**

There are various medical conditions which can result in a person needing a PEG tube. It may be suggested if someone is finding it difficult to swallow food and drinks and as a result they may find it difficult to take in enough to meet their nutritional requirements. This could mean they may become malnourished or dehydrated which could be damaging to their health and well being.

All nutrition and fluid can be given via a PEG tube, so if eating and drinking become a struggle or mealtimes are taking a long time someone's nutritional needs can be met by providing them with a special liquid feed. This can allow more time for other things and you can still eat small amounts for pleasure if you wish.

A PEG can be used for

- ◆ Full feeding, i.e. providing all the nutrients and fluid you need
  
- ◆ Or just to top up the food and drink being taken by mouth to ensure you are getting all the nutrients and fluid you need.

### **Are there any complications?**

Having a PEG fitted is a relatively safe procedure but as with every medical procedure there is some risk involved and some complications can occur.

### **What feed will I receive?**

- ◆ You will receive a ready made feed which contains all the essential nutrients you need on a daily basis.
- ◆ Before you go home your dietitian will discuss your feeding regimen with you and how to look after your tube and make sure you and your carer are fully trained.
- ◆ The method of feeding will be adapted to suit your lifestyle at home. Feed can either be given as small meals or boluses throughout the day or more slowly overnight.
- ◆ Your doctor will provide prescriptions for your feed and it will be delivered to your home along with any other equipment you may need..

### **What can I put down the PEG?**

In general the only things that should go down the PEG are

- ◆ Your feed
- ◆ Water
- ◆ Medicines

### **How long will the tube last?**

This varies from patient to patient but they can last several years.

### **Is the tube likely to fall out?**

There is a “device” in the stomach that is attached to the tube to hold it in place and should prevent it falling out.

### **Is it possible to take a bath or shower?**

- ◆ You can take a shower the day after the PEG is placed, but it needs to be covered with a waterproof dressing.
- ◆ After the PEG site is healed (about 2-3 weeks) you are able to take a bath but you must make sure you dry around the site thoroughly.

### **Finally**

Please remember this leaflet is intended to provide you with some basic advice about PEG feeding so you know a little about what is involved and what it can and cannot do. It is to help you to think about whether or not you may wish to consider having one should the need arise in the future. It may have made you think of more questions you would like answered, if that’s the case and you or anyone else would like to discuss this further please feel free to contact 0151 2983298.

Huntington's Disease Association

Standards of Care

# End of Life



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## Introduction

### Options at the End of Life

#### What are the options for someone in the later stages of HD?

The options for a person with later stages of HD should be the same as any other person with a life-limiting disease. Depending on the area where someone lives, the options should be:

- At home – with support from district nurses and hospice-at-home services where appropriate / available.
- Residential Home – again with support from district nurses and hospice-at-home services where appropriate / available
- Nursing home – nursing homes should have access to a Macmillan Team who can advise on symptom control or should be able to access the neurology team at the hospital
- Hospice – ‘Hospice Information’ provides information on hospice care and a ‘find a hospice / palliative care service’ facility (see resources)
- Hospital

A HDA Fact sheet on choosing a care home is available (see resources)

#### How should the decision be made as to which is the most appropriate option for this person?

The decision should be person-centred, and take into account the individual's personal wishes and circumstances. The decision-making process should involve the multi-disciplinary team.

The *Preferred Priorities for Care document* (see resources) (formally known as Preferred Place of Care) is an Advance Care Plan, where people can document their preferences and priorities for care at the end of life.

### Ascertaining a person's wishes

#### How should a person with HD's wishes (and those of their family) be identified?

A discussion about a person's wishes, and any decisions that need to be made, should happen as early as possible in consultation with their family. This ensures that the person with HD has their own input and decisions do not have to be made by others in crisis / best interest scenarios when the individual has lost capacity.

These decisions should be reviewed regularly and should be fully recorded. These records should be available to all who are influential in providing care. The best place to keep this information may be with the person with HD.

#### What issues should be discussed with the person with HD (and their family)? and who should be involved?

Issues that should be discussed include:

- Likes and dislikes (e.g., food stuffs, music, television etc.)
- Previous experience (e.g., family history, what happened and how did they feel about this?)
- Disease projection and what to expect
- Place of care / place of death
- Resuscitation and ventilation
- Artificial feeding - Further information about artificial feeding, and how this decision should be made can be found in the *diet, swallowing and communication* standard, and readers are referred to this for more information
- Organ donation - Information about brain donation is available from the HDA Head Office. This includes an information sheet for patients/relatives, guidance notes and a consent form. If someone contacts you after a person dies then speed is essential as to be of any use the brain must be removed quickly. In this instance it is better to contact the brain bank directly - London Neurodegenerative Diseases Brain Bank, PO Box 65, Institute of Psychiatry, 4 Windsor Walk, London, SE5 8AF. T:020 7848 0290 E: brainbank@iop.kcl.ac.uk
- Lasting Power of Attorney (LPA) – An LPA is a legal document allowing an individual to appoint someone to make decisions regarding ‘property and affairs’ and / or ‘personal welfare’ on their behalf. A LPA can only be used after it has been registered with the Office of the Public Guardian (OPG). More information and guidance booklet are available from the OPG website (see resources)

A HDA Fact Sheet on ‘Huntington’s Disease and the law’ is also available (see resources)

- Advanced Decisions to refuse treatment (ADRT) – In October 2007 the ADRT replaced ‘living wills and advanced decisions’; they are increasingly recognised with medical practitioners. Discussions should take place about preparing an ADRT, and the person should be supported in doing this if they wish. N.b. once a ‘personal welfare’ LPA has been registered with the OPG the ADRT becomes invalid. The HDA have a fact sheet on Advanced directives (see resources) and are in the process of creating a model document (please contact the HDA Head Office for a copy), the NHS provide an on-line training programme in ADRT (See resources)

All those involved in a person’s care should be aware of their wishes; this can be difficult if the person is admitted to emergency care for an acute event. The HDA have a ‘key facts’ leaflet which can be useful in this event (see appendices)

In an ideal world the discussions should be led by GP / neurologist / psychiatrist or multidisciplinary team in consultation with person with HD and family members and then documented. This may

take several months in order that the person has informed consent. These discussions may involve talking about some very difficult issues, which requires a lot of sensitivity, knowledge and expertise.

### Management of Symptoms

#### How can the needs of someone with HD in the later stages best be assessed (and re-assessed)?

Caring for someone with HD in the end stages of life should be no different than caring for anyone else at the end stage of life, and the issues should be familiar to members of primary care teams / other palliative care services.

Assessment of a person's needs may be difficult if communication and / or cognition are impaired, as is often the case in the later stages of HD. The onus is then on carers / professionals to work through a checklist of symptoms and get to know the person as an individual. Visual observation and simple communication systems (e.g., touching of hand to indicate yes / no) may help in these situations. Some pain scales e.g., *Abbey Pain Scale* (see resources) offer a checklist of non-verbal pain indicators. A detailed plan of care can help in these situations.

#### What symptoms may cause difficulty in the later stages of HD?

The following may cause difficulty in later stages of HD:

- Changing sensitivity to pain and temperature
- Aspiration
- Infections
- Loss of swallow reflex
- Malnutrition / dehydration
- Weight loss
- Reduced mobility

Each symptom should be assessed individually and treated with the same palliative care principles as any other person at the end of life. It is important to recognise that not all symptoms will be due to HD, and to be aware of any changes that are identified being caused by something else.

#### How should these best be managed?

Symptoms in the later stages of HD should be managed holistically with input from palliative care physicians / nurses. Good communication with the family / patient and with other members of staff within the team caring for the person with HD is also important. Further information about managing these symptoms can be found in the *diet, swallowing and communication* standard, the HDA book – 'Caregivers Guide for Advanced Stage HD' and in the Scottish Huntington's Association *Care Pathway* (see resources).

### What resources are available to help manage these symptoms / issues?

Palliative care teams from a local hospice should be available to GP's / other physicians as a resource when managing patient's at the end of life (e.g., hospice at home services, Macmillan or Marie Curie nurses). Many areas have equipment banks where equipment can be loaned e.g., syringe drivers.

### Family Issues

#### What are the family issues that should be taken into consideration in the care of people with HD?

The hereditary nature of HD often means that there is repeated loss in families. Bereavement / grief work is a particularly important area. Care homes may have members of the same family within it and special attention should be paid to this. The carer / other family members may also be at-risk from developing HD, or have tested positive for the HD gene and this may affect their ability to deal with the situation.

#### How should family members be best supported?

Family members should be supported from diagnosis, through death and beyond by the multi-disciplinary team. Local bereavement support is often available through hospices or other organisations e.g. Cruse Bereavement Care (see resources)

### Staff training / Support

#### What training should staff receive on HD / palliative care?

Relevant training in HD and palliative care should be offered to staff that require it. Examples include: communication skills training, Macmillan Foundations in Palliative Care, HD Awareness training, and training in the use of end of life tools previously mentioned. The Regional Care Adviser for the Huntington's Disease Association can offer training on HD. Local hospices often have training courses for external staff.

#### What support should be available for staff caring for someone with HD?

It is important to recognise that staff caring for someone over a period of time will need help and support during this time, and after the person dies. Those who are caring for someone with HD also require support to do this. For example, clinical supervision, access to education / training on HD and end of life care.

## Resources in relation to end of life care

### HDA Resources

Factsheet 12 Huntington's Disease and the Law

Factsheet 16 Checklist for choosing a care home

Factsheet 17 Advance Directive or "Living Will"

Pollard, J (ed.) (2003) Caregivers Guide for Advanced Stage Huntington's Disease. Huntington's Disease Association

### Journal Articles / Books

Addington-Hall, J.M. & Higginson, I.J.(ed) (2001) Palliative Care for Non-Cancer Patients Oxford, Oxford University Press.

British Medical Association (2007) Withholding and Withdrawing Life-prolonging Medical Treatment. Guidance for decision making (3<sup>rd</sup> Ed) Oxford, Blackwell Publishing Ltd.

Byrne, J and Beety, D (2007) Tools to Improve End-of-Life Care for Neurological Patients. British Journal of Neuroscience Nursing 3 (5) 190-193

Campbell, C. & Partridge, R. (2007) Artificial Nutrition and Hydration. Guidance in end of life care for adults. London, The National Council for Palliative Care.

Doyle, D. et al (ed) (2005) Oxford Textbook of Palliative Medicine Oxford, Oxford University Press.

Jeffrey, D. (2006) Patient-centred Ethics and Communication at the End of Life Oxford, Radcliffe Publishing.

King, N. (2005) Palliative Care Management of a child with juvenile onset Huntington's disease International Journal of Palliative Nursing Vol 11, No 6, 278-283

Wasson, K (2000) Ethical arguments for providing palliative care to non-cancer patients International Journal of Palliative Nursing Vol 6, No 2, 66-70

### Internet Resources

- **Hospice information** is a joint venture between St. Christopher's Hospice and Help the Hospices. It brings together the experience and established reputation for high quality of the Hospice Information Service at St. Christopher's and the national remit and innovative information developments of Help the Hospices. Includes 'find a hospice / palliative care service. <http://www.hospiceinformation.info/index.asp>

- **NHS End of Life Care Programme (EoLC):** The NHS End of life Care Programme (EoLC) was set up to improve care at the end of life for all wherever they live. This site aims to support the programme by sharing good practice, resources and information. <http://www.endoflifecare.nhs.uk/eolc>
- **The Gold Standards Framework (GSF):** The GSF is a systematic evidence based approach to optimising the care for patients nearing the end of life in the community. It is concerned with helping people to live well until the end of life and includes care in the final year of life for people with any end stage illness. <http://www.goldstandardsframework.nhs.uk/>
- **Liverpool Care Pathway for the Dying Patient (LCP):** The LCP Framework is a continuous quality improvement framework for care of the dying irrespective of diagnosis or place of death. [http://www.mcpcil.org.uk/liverpool\\_care\\_pathway](http://www.mcpcil.org.uk/liverpool_care_pathway)
- **Preferred Priorities for Care (PPC):** The PPC is intended to be a patient-held record that will follow the patient through their path of care into the variety of differing health and social care settings. Guidance reference sheets for both the patient and carer and staff are available explaining the use of the PPC. <http://www.endoflifecareforadults.nhs.uk/eolc/eolc/current/CS310.htm>
- **Advance Decisions to Refuse Treatment (ADRT) Training Programme:** This is an on-line training programme for professionals. The aims are for the professional to understand the legal context of the ADRT, to understand the reason why people may wish to make an ADRT and to help support patients in the process of making an ADRT. <http://www.adrtnhs.co.uk/>
- **Cruse Bereavement Care** - Cruse Bereavement Care exists to promote the well-being of bereaved people and to enable anyone bereaved by death to understand their grief and cope with their loss. The organisation provides support and offers information, advice, education and training services. <http://www.crusebereavementcare.org.uk/>
- **Department for Constitutional Affairs (DCA):** The DCA has a range of literature available on Lasting Power of Attorneys. They have a leaflet 'making decisions about your health, welfare and finances....who decides when you can't' this is available in a number of languages. Also available are the following booklets:
  - For people who may be unable to make some decisions for themselves/ who wish to plan ahead for the future
  - For family, friends and unpaid carers
  - For people who work in health and social care
  - For advice workers
  - The Independent Mental Capacity Advocate (IMCA) service

<http://www.dca.gov.uk/legal-policy/mental-capacity/publications.htm>

- **Office of the Public Guardian (OPG):** The OPG helps to protect people who lack capacity, it's role includes setting up and managing a register of LPA's. The following guidance on LPA is available to download
  - A guide for people who want to make a personal welfare Lasting Power of Attorney
  - A guide for people who want to make a property and affairs Lasting Power of Attorney
  - A guide for people taking on the role of personal welfare Attorney under a Lasting Power of Attorney
  - A guide for people taking on the role of property and affairs Attorney under a Lasting Power of Attorney
  - A guide for Certificate providers and Witnesses
  - A guide to registering a Lasting Power of Attorney
  - Examples of what a registered Lasting Power of Attorney will look like

<http://www.publicguardian.gov.uk/index.htm>

- **Abbey Pain Scale (The Abbey):** The Abbey Pain Scale is an Australian tool developed to measure severity of pain in people with late-stage dementia  
<http://www.cityofhope.org/prc/Review%20of%20Tools%20for%20Pain%20Assessment/Abbey%20Text.htm>
- **Scottish Huntington's Care Pathway.** <http://www.hdscotland.org/>
- **The Open University:** The Open University Learning Space gives free access to course materials from the Open University. Among other topics the 'health and lifestyle' unit offers the following:
  - Ageing and Disability: Transitions into residential care
  - Moral and Ethical Principles in End of Life Care
  - Living with death and dying

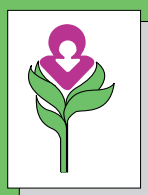
<http://openlearn.ac.uk/>

- **"Palliative Initiatives in Neurological Care" (The PINC Project):** Sue Ryder Care have piloted three nationally recognised end of life Care Tools – *Gold Standards Framework in Care Homes*, *Integrated Care of the Dying Pathway* and *Preferred Place of Care (now Preferred Priorities for Care)*. The outcomes will be published at the end of June and will be available on the Sue Ryder Care website. <http://www.suerydercare.org/>

# Appendices

Appendix 1\_ Key Information about Huntington's Disease – A quick reference guide for hospital staff

  
More information  
is available from the  
**Huntington's  
Disease  
Association**  
by telephone  
+44 (0) 151 298 3298  
on our website  
www.hda.org.uk  
or by email  
info@hda.org.uk



Neurosupport Centre,  
Norton Street,  
Liverpool. L3 8LR  
Tel: +44 (0) 151 298 3298  
Fax: +44 (0) 151 298 9440  
Email: info@hda.org.uk  
Website: www.hda.org.uk  
Regd. Charity No. 296453

## Key information about **Huntington's Disease**

It's **hereditary** - children are at risk of developing the condition.  
and **progressive** - **there is no cure but much can be done to control symptoms.**  
People develop symptoms covering motor, cognitive and emotional changes and these can vary considerably among individuals who have Huntington's Disease.

### Communication difficulties

**There can be a delayed response to answering questions** - ask closed questions, one at a time. Some individuals may use communication aids, such as Word Boards etc. Seek advice from SALT and carers.

### Swallowing Difficulties

- **Danger of choking** - patients need assistance and supervision with feeding and drinking.
- **Positioning** - patients need to be sat upright for oral intake.
- **Minimal distraction** whilst eating.
- **Consistency of food** needs to be considered - seek advice from speech and language therapist, dietician and family carers.
- **Small frequent meals** - don't mix textures.
- Patients with Huntington's Disease need **more calories than normal** - consider supplements- It is essential to maintain body weight.

### Alternative Feeding

If a patient has a PEG fitted, information and advice on usage and care should be sought from the dietician and family members/carers.

**If the patient has been admitted with swallowing, feeding or aspiration difficulties, accurate information and wishes should be sought from family members/carers and professionals involved in their care.**

Any decision about fitting a PEG tube needs multi-disciplinary consideration, preferably from professionals previously involved with the patient. It is essential that family members are consulted and patients' previous wishes explored.

### Cognitive impairment

**Understanding is often maintained in the late stages.** Non-verbal communication may be misleading owing to patients' limited facial expressions.

### Behavioural presentation

Organic brain changes result in the fact that -

- people with Huntington's Disease cannot adapt their behaviour - **you must adapt yours.**
- **routine is important** - people with Huntington's Disease can't wait for anything.
- emotional states may include anxiety, irritability, agitation, apathy and depression. Further advice may be sought from psychiatric services.
- Any **sudden changes in behaviour** may relate to other illnesses, pain, hunger or thirst.

### Movement disorder

Involuntary movements may be problematic and safety is important. Rigidity may develop in late stages and the patient may develop associated pain.

### Medication

**Generally any medication previously prescribed should not be stopped suddenly** but needs gradual reduction. It is important to obtain information about, and the reason for, any currently prescribed medication.



## NOTES

## NOTES



# HUNTINGTON'S DISEASE ASSOCIATION

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Tel/Fax: 0151 298 3298 E-mail: [info@hda.org.uk](mailto:info@hda.org.uk)  
Web: [www.hda.org.uk](http://www.hda.org.uk)

Registered Charity No. 296453