


That

makes

sense

now



Decoding the symptoms
of Huntington's disease



Huntington's
Disease
Association

Interpreting the varied symptoms of Huntington's disease can be difficult

Huntington's disease (HD) symptoms can vary widely between individuals and sometimes present in ways which are not immediately clear to the healthcare professional. Occasionally the symptoms are mistaken for other conditions such as Parkinson's disease or Alzheimer's dementia, so being able to recognise the unique behaviours and physical presentations that accompany the disease can help us to take the optimal route to improving care.

Consultant Neuropsychiatrist, St Andrew's Healthcare

Huntington's disease is a rare and complex illness that can offer care givers many challenges. Its unique combination of mental health problems, physical impairment and cognitive 'slowing down' often means that behaviours and symptoms can be misinterpreted. By having an understanding of these it can help us improve the care that we give and thereby improve the quality of life for people with HD and their families.

Cath Stanley, Chief Executive, Huntington's Disease Association



It's not the person

That

behaves that way, it is the disease

There are many aspects that should be considered

It **makes**

it easier to anticipate their needs

*I used to find it hard
to make*

sense *of his behaviours*

I had never looked after someone with HD before

*When she wants something,
she wants it **now***

I used to think that too many calories was a bad thing

Making some simple changes has transformed their quality of life

What is Huntington's disease (HD)?

HD is a relatively rare inherited genetic disease and you may not yet have come across a person who has been diagnosed with HD or is displaying symptoms. It is caused by a faulty gene on chromosome 4, producing a protein called Huntingtin which is thought to increase the rate of cell death in the brain, and affects about 8,000 people in the UK.

HD affects the central nervous system and usually develops when individuals are in their thirties to fifties, or in the case of Juvenile HD, those under the age of 21. Each child with a parent with HD has a 50:50 chance of inheriting the condition and a test can be performed to determine whether the faulty gene is present.

There is no cure for Huntington's disease.

"It's a lot of detective work"

Because HD presents in ways which can be confusing at times for carers it is important to unravel the many 'mixed messages' and layers of disorder so you can truly understand how best to support the patient. Recognising the unique behaviours and symptoms of HD can guide us to take a different approach to treatment that improves care and preserves the dignity of the individual.

"It is, however, the disease and not the person who is at fault. People with HD are not being deliberately thoughtless, awkward and uncaring. There are no easy answers... however, understanding why people with HD behave in the way that they do is important since it may provide clues."

Julie S. Snowden, Understanding challenging behaviour in Huntington's disease, 2002.

Supporting people with HD therefore requires an investigative approach to understand the root cause of individual behaviours and why they may be causing specific responses.



Understanding the effects of HD supporting healthcare professionals

The effects of HD are wide ranging and **change how people think, feel, speak, move, swallow and eat**. It presents typically at a time in life when most people may be establishing families and careers yet those with the disease are suddenly faced with the emotional impact of impairment and living with a disease for which there is currently no cure.

These effects can be broadly grouped in to three key elements of change, all of which affect how people with HD behave and influence how we should consider their needs:

The three elements of HD deficit can be summarised as:



Physical

the motor functions, particularly those controlling movement and eating

Physical symptoms of HD including voluntary and involuntary movement are, perhaps, the most obvious. However there are many other symptoms, from swallowing and speech difficulty to muscle rigidity, that require specialist input and care.



Cognitive

the processes that manage how people think, plan and interact

The cognitive disorders may be less obvious in early stages of the disease and can often be confused with other conditions, or simply attributed to forgetfulness, laziness or a general lack of awareness. As the disease progresses these become more pronounced and disabling.



Emotional

the mood and behavioural changes that affect /reflect how people feel

Emotional and psychological symptoms develop as the disease progresses (and even before symptoms become evident) as people come to terms with the prospect and results of diagnosis. Living day to day with HD can wear individuals down as the debilitating effects of the disease directly impacts on mood and behaviour.

HD symptoms and implications



Physical

Physical symptoms of HD include:

- **Chorea:** Greek for 'dancing', a movement disorder causing 'jerky' uncontrollable actions. The person may over-compensate for this with big bursts of movement.
- **Dysphagia:** Difficulty in swallowing and sometimes bringing food and drink back up, increasing the risk of choking. Dieticians can recommend graded foods.
- **Rigidity:** More often at latter stages making fine motor control, such as holding objects, difficult.
- **'A misleading disguise':** Changes in appearance caused by weakness in facial and body muscle tone may appear as boredom or anger, and poor posture may look like 'attitude'.
- **Reduced volume control:** Muscle deterioration in the throat means the person may switch between whispering and shouting for no obvious reason, making it difficult to gauge mood and needs.
- **Slurred speech:** Muscle deterioration can also change the tone and coherence of speech, and is sometimes mistaken for drunkenness.
- **Weight loss:** An on-going challenge as changes in metabolism, combined with their constant movement, mean that people with HD burn more energy, so require a high-calorie diet.
- **Incontinence:** Understandably a cause of distress, and requires support from urology teams.
- **Lowered immune system:** Underactive immune protection makes the person more prone to illness, and requires careful management.



"Because balance becomes impaired the risk of falls is much greater. Consider how best to safely transfer patients in and out of beds, chairs etc, and pay close attention to walking."

Carer



Cognitive

The cognitive disorders include:

- **Difficulty planning ahead:** A deteriorating ability to plan tasks and stick to schedules.
- **Difficulty multi-tasking:** The person may become fixed on a task or activity, to the detriment of other things such as hygiene.
- **A need for routine:** A preference for scheduled times for eating, bathing, dressing etc where changes can make the person obstinate. Inform and plan changes with the person.
- **Lack of initiation:** Difficulty getting started with tasks and activities, such as maintaining hygiene.
- **Poor memory and concentration:** Can affect short and long-term memory, requiring prompts.
- **Repetition:** Repeating the same questions and getting 'stuck' on a subject, make it difficult to move topics on.
- **Inability to read facial expressions:** It may appear that the person is oblivious to the mood of others, but they have difficulty reading other people's faces.
- **Extra processing time required:** It may take longer, sometimes minutes, for the person to respond to a question or request.
- **Lack of insight or awareness:** A general inability to gauge their actions as the disease progresses.



Emotional

The emotional and psychological symptoms include:

- **Mood and behaviour changes:** Family may notice the person becoming moody and irritable more frequently and for longer periods of time.
- **Impulsivity:** Changes in the brain may make the person more impulsive in their speech and actions.
- **Unable to wait:** This is not because the person is selfish or impatient, but a direct impact of the disease on the brain.
- **Communication challenges:** "I know exactly what I am saying... why can't you understand me?" Difficulty relaying feelings, information and requests can lead to misunderstandings with carers.
- **Frustration:** Cognitive changes can cause frustration which can occasionally lead to temper outbursts.
- **Apathy:** People who have always been active may suddenly find it difficult to motivate themselves, and may just prefer to watch tv.
- **Depression:** Common throughout all phases of the disease, and important to liaise with specialist services, such as GP and psychiatry.
- **Repetitive loss and bereavement:** The person faces the constant re-realisation that their present, future, independence, family-life etc will change.
- **Irrational and disinhibited behaviours:** Some people may develop obsessions (e.g around objects or people), and displace some of the social norms that govern our lives, such as undressing in public.
- **Social isolation:** Whether self-imposed or as a result of the impact on friends and family.



"I used to have a great job and an active lifestyle. Just because I have HD doesn't mean I have forgotten how to have fun."

Ian, living with HD



The importance of specialist care

Including specialists in care planning from an early stage can be the difference between a high or a compromised quality of life for someone with HD.



"Expert medication management will help the symptoms of mental illness that frequently occur alongside the movement disorder."

Neuropsychiatrist



Managing Cognitive Deficits:

- baseline assessments of cognitive function
- creating bespoke strategies for care
- planning the wrap-around specialist therapies and treatment
- occupational therapy to support daily living
- formulation-guided interventions
- graded access to community
- assessment of personal safety awareness
- guidelines for continuing support as ability decreases



"Provide a comprehensive assessment of cognitive function to help understand the limits of a person's ability to think, plan or remember."

Clinical Psychologist



Neuropsychiatry and Neuropsychology:

- recognising, assessing and managing the HD symptoms
- tailoring nursing and specialist care to each individual
- creating a timetable of activities / outings to support treatment
- supporting emotional needs as the disease progresses
- 'Life story' work to truly understand the person's likes and dislikes
- mindfulness meditation
- emotional management guidelines for patients and staff
- adjustment and commitment therapy
- enhancing opportunities for family visits
- supporting interpersonal skills / relationships
- art and music therapy

MDT



Managing Physical / Motor deficits

- physiotherapy
- speech and language therapies
- management of dysphagia through individual dietary assessments
- dysarthria therapy to support speech and communication
- access to complex medical investigations such as videofluoroscopy or PEG fitting
- review of equipment and seating
- access to specialist equipment and assistive technology
- involvement of community specialists such as GP and podiatry services
- OT focus on movement, mobility and posture



"Regular swallowing assessment can guide the grade of diet and consistency of fluids that can be safely managed to reduce risks of choking and aspiration."

Speech & Language Therapist

Because I have HD...

You may find it helpful to share this cut-out page with other care-givers, or include it in the person's care notes.

Hi,

I have Huntington's disease (HD) which is a brain disease that affects how I think, feel and act, so I thought I would share a few things that might help you to help me.

Because I have HD it means:

- I like my own routine
- I do one thing at a time
- You need to get my attention first and then tell me what you want
- You need to give me time to answer – don't repeat what you said or put it another way as this makes it difficult for me to answer
- It takes a lot of effort to speak – listen to what I say
- I sometimes cannot control how loud (or quiet) I am
- I don't know how to wait! If I need something I really need it now
- I lose weight quickly so need lots of little snacks and drinks
- I find it hard to control my body, so am constantly moving
- 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.



"The risk of choking is very high, particularly as some people eat too fast and forget to chew. Find a quiet area for them to eat in and always keep an eye on them."

Carer

You can help me by considering:

- How can you best minimise communication difficulties?
- What diet plans are in place: What food, how often and how much?
- How you can help me to eat
- Do you know how to prevent dysphagia or choking?
- How can you support emotional problems I may be facing?
- Do you have a programme for managing continence and know which aids to use?
- How can you best optimise my mobility and the associated risks?
- Do you have a plan to manage my medication, and understand what it is?
- Do you understand my sleep patterns, and have you planned care around them?



"Make sure that nutritional needs are met and a high calorie diet is maintained. Think of people with HD as always being hungry, so don't be afraid to offer double portions! The prescription of specialist supplements can also help to slow weight loss and prevent malnutrition."

Dietician



About the Huntington's Disease Association (HDA)

The HDA was established in 1971 as a specialist service to help people and families affected by Huntington's disease (HD). The HDA is a national charity, working to improve the lives of people with HD, and raise awareness among the general public and allied health and social care professionals.

Their award winning Advisory Service is vital in providing the best package of support for families. Advisers offer care to the carers (often spouses and children) and play a pivotal role in keeping the family together. This support is paramount as people who are symptomatic of HD exhibit a wide range of problems including severe depression, violent behaviour and difficulties in swallowing. Advisers also act as an advocate, providing specialist information and detailed training to health and social care professionals, and a large part of their role is to discuss end of life care openly and provide bereavement support to the family and carers when an individual dies.

The helpline team offers a confidential service providing support to anyone affected by Huntington's disease. They can answer questions on medical issues including symptoms and treatments, employment and benefits, emotional support and signposting to other agencies.

To speak with a member of the team please call

0151 331 5444

or visit the website at hda.org.uk



**Huntington's
Disease
Association**

"Our specialist Huntington's disease adviser provided excellent advice and guidance to help me liaise with social services and the NHS. Her experience made a difficult time much easier to cope with."

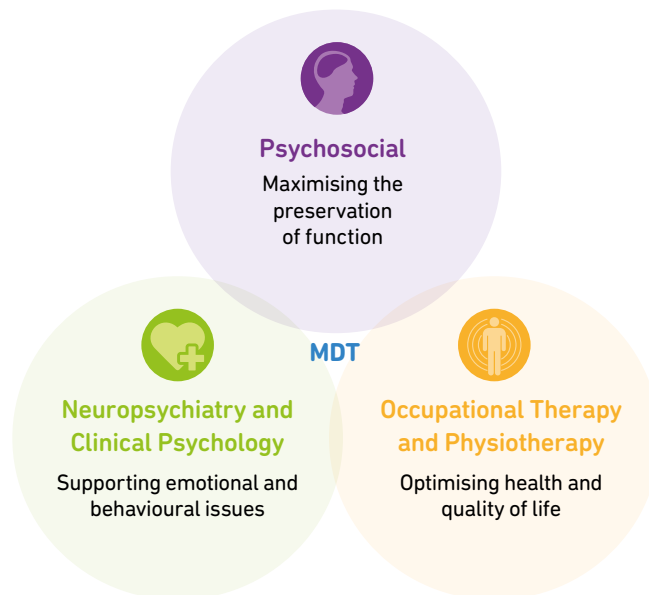
Person affected by Huntington's disease, 2015

“That makes sense now”

At St Andrew's we are expert at making sense of the physical, emotional and psychological impact of living with HD. Our specialist teams have developed pathways and therapies which deliver person-centred care tailored to the needs of the individual, through all stages of the disease.

We are a mental health charity and have devoted over 175 years to supporting patients with neuropsychiatric needs. We focus on maximising functional ability and quality of life by delivering holistic management of psychiatric, cognitive and motor deficits. Our environments meet the unique physical needs of patients, and our ethos reflects our warm, positive and empathic approach.

Each patient is surrounded by a comprehensive group of in-patient and community specialists to ensure they receive the most appropriate, tailored care.



We hope that you have found this a useful guide to providing the best possible care for people with HD. If you would like to find out more about HD and how to support people living with HD (and their family and carers) then you can contact the useful links below.

Useful links:

- Huntington's Disease Association (HDA), England & Wales, **0151 331 5444**, email info@hda.org.uk, website www.hda.org.uk
- Scottish Huntington's Association (SHA), **0141 848 0308**, email sha-admin@hdscotland.org, website www.hdscotland.org
- European Huntington's Disease Network (EHDN), website www.euro-hd.net
- HDBuzz, website www.en.hdbuzz.net
- The HD Lighthouse, website www.hdtf.org
- NHS Choices, website www.nhs.uk/conditions/huntingtons-disease

The importance of waiting

James Pollard in his book *Hurry up and Wait* emphasises how the difficulty people with HD have in controlling their impulses is caused by changes in the brain, and not because the person is selfish, impatient or aggressive.

The result is that people with HD simply cannot wait... and if they want something they want it now!

It is important to **wait for the person with HD to understand what is going on**, consider their thoughts and communicate their response. This can sometimes take time and the carer can provide visual prompts or tools to help them process information and communicate their needs.

On the other hand people with HD need immediate resolution or action, such as "I want some food now". Try to always **respond immediately**, and whenever possible predict the needs of the patient. The more time you spend understanding their likes,

dislikes, needs and preferences, the more likely you are to be able to anticipate what the patient wants and reduce their frustration.

Whether you are a family member or carer of someone with HD you will recognise many of the symptoms highlighted in this booklet. However for most healthcare professionals this will be the first time you will have come across HD so we hope this provides some clues to help you to understand the symptoms and improve care for those living with the disease.

Getting in touch



For more information about our HD service or to make a referral:

t: 0800 434 6690 (text relay calls welcome)

e: enquiries@standrew.co.uk

w: stah.org



For more information about HD or to speak with an HDA adviser:

t: 0151 331 5444

e: info@hda.org.uk

w: hda.org.uk