



**Huntington's
Disease
Association**

**Huntington's disease:
A guide for social
workers**

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Introduction

Huntington's is a relatively rare disease. About one person in 10,000 has Huntington's disease. Many social workers may have never encountered someone with the disease before. This leaflet contains important information to help social care workers understand more about Huntington's disease and the challenges people affected may be facing.



One of the biggest challenges social care workers will face, in assessing a person with Huntington's needs, is that they often lack insight into their own abilities and will often say "I can manage, I'm fine" when it is clear they are not. It is important to bear this in mind and take advice from family and other healthcare workers that know the person.



It is important to remember that it may take longer to assess a person with Huntington's disease due to communication difficulties and slow cognitive processing of information. It is good practice to not take their first response to questions at face value.



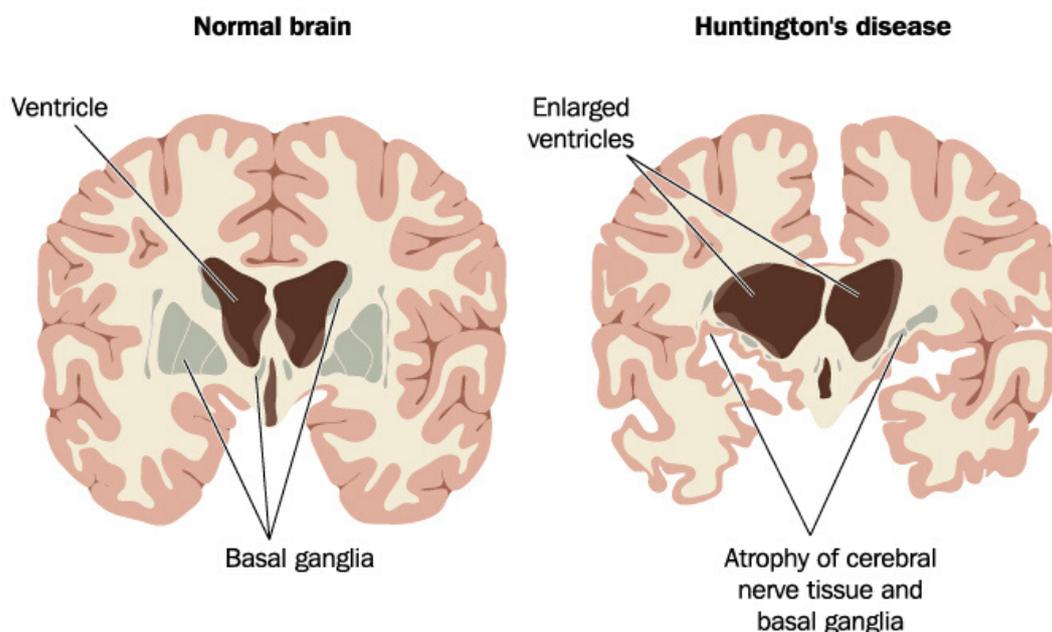
What is Huntington's disease?

Huntington's disease is a disease of the central nervous system. It is caused by a faulty gene and is passed down through families. If a person inherits the faulty gene, they will develop the disease.

Every child conceived naturally to a parent who carries the Huntington's gene has a 50% chance of inheriting it.

Around 8000 people in the UK have Huntington's disease and it affects both men and women.

Huntington's disease causes significant degeneration of the part of the brain known as the basal ganglia. This is the part of the brain that helps to co-ordinate movements, cognition and emotion. The following image shows the change that happens in the brain of someone with the disease. As you can see, the most significant damage is in the basal ganglia but there are also more subtle changes occurring in other parts of the brain. The brain itself is slightly smaller and weighs less than normal.



Huntington's disease changes the whole person - body, mind and behaviour causing changes in their movement, the way they act and their mental well-being. As the disease progresses, the person will lose their ability to walk, talk, think clearly, swallow and control their movement. The disease can cause major depression, obsessive behaviour, paranoia, psychosis and thoughts of suicide.

Symptoms usually begin between the ages of 30 and 50, but can start at any age. Children and young people under 20 years old who show symptoms early, have Juvenile Huntington's disease - an even rarer form of the disease.

There is no cure yet. But, there are treatments available to manage symptoms.



10 things you need to know about Huntington's

- 1 It is a genetic disease inherited from a parent.
- 2 It is caused by a faulty gene that makes areas of the person's brain function inefficiently.
- 3 It usually progresses over a period of 15 to 20 years, but can progress faster or slower.
- 4 It affects thinking, speaking, swallowing, movement and behaviour.
- 5 People with Huntington's can appear bored, disinterested or angry when they're not.
- 6 Symptoms often begin when most people have begun a career, started a family and have been doing well in life.
- 7 Most likely the person has watched a parent suffer and is worried that each child they have has a 50% chance of developing Huntington's too.
- 8 There is no cure yet.
- 9 There is treatment for control of mood and movement.
- 10 With support and co-ordinated care, the person with Huntington's can keep their independence for as long as possible and live a better life.



Behaviour, thinking and mood

Due to deterioration in the brain, people with Huntington's disease find self-monitoring difficult and have a lack of awareness that the disease is affecting them physically, emotionally and cognitively. The changes to the brain also make it hard for those affected to envision the future and they can be reluctant to consider anything new. This behaviour can be difficult to modify as the person with Huntington's may accept they have the disease but not that they are showing symptoms. This unawareness can become dangerous, resulting in the person with Huntington's putting themselves or others at risk. There are a number of strategies you can use to help.



Useful strategies to try

- **Get to know the person** - building a relationship with the person with Huntington's is important and may take time. You may need to assess the person over multiple visits and it may take a while for the person to agree to any help offered.
- **Visit at home** - this will give you a better idea of the situation as people with Huntington's will often say they are 'fine' on the phone when they are not.
- **Use the person's support network for information** - seek information and advice from family, friends and other professionals who know the person well. Consider joint visits.
- **Carer support** - listen and support those who are caring for the person with Huntington's disease.
- **Take your time** - a person's ability to process information is slowed in Huntington's disease. Make sure you give the person time to understand, process and respond.

- **Try different methods of contact** - Huntington's affects the person's ability to plan, organise and initiate activities. They may not always respond to calls, letters or visits. Don't take this as refusing contact. Try contacting the person at different time of the day and using different methods.
- **Reassure about the future** - people with Huntington's find change and picturing a different future (perhaps a life with help in place) difficult. Change can make them anxious and reluctant to accept help. Encourage the person to try something out and see how it works.
- **Start small** - starting with small amounts of help and support can mean the person gets used to having and accepting it. Even if someone is physically able to do something, they may still need a lot of support with prompting / monitoring.
- **Don't make insight the main goal** - trying to tell someone they have Huntington's when they are in denial will likely make engagement more difficult. If you are aiming to get the person to accept help or manage the risks associated with their symptoms, it is more practical to address the issues at hand rather than forcing them to confront the fact they have Huntington's disease.
- **Be flexible and creative** - where possible, try to work on what the person will accept help with. If they will accept help with cleaning but not personal care, begin with cleaning support.
- **Seeing it from another perspective** - a person with Huntington's may not accept help for themselves but may accept help for their loved one. For example, they may be able to see that their loved one is overworked and could do with some help with cleaning.
- **Keep trying** - most people with Huntington's will accept help eventually but you may need to wait until the right moment arises.

"I'm fine, I don't need any help"

This is undoubtedly the most challenging aspect of Huntington's disease. As the person with Huntington's is unable to self-appraise or have insight into their own abilities, they may frequently insist they are "fine" when they are clearly not managing even basic needs of cleanliness and diet. Please do not give up on someone who says this. It may take several visits and time to build a relationship before they are willing to accept even the smallest amount of help.

Most people with Huntington's disease are anxious to cling on to independence, but with time and a calm approach most will allow some degree of help. Once support is agreed and in place, there may still be some reluctance by the person to accept help. There are some strategies that can help.



Strategies to consider

- **Routine is important** - once a care plan has been agreed, a set routine and consistency between carers is important for people with Huntington's disease.
- **Reiterate the plan** - you might consider encouraging the person to use wall planners, reminders on their phone, verbal reminders that the person with Huntington's and their carers can refer to about any agreed care and support.
- **Listen and try to understand** - consider why the person is reluctant to do something. For example, do they not want to shower because they feel unsteady when they're in the shower?
- **Consistency** - people with Huntington's find change difficult. Carers arriving late or different carers arriving each day is likely to cause upset and distress. If there are particular carers the person responds to best, look at the methods they use and try to replicate this across the team.

- **Time is important** - people with Huntington's need time. If the person says no, a carer may need to come back a bit later and ask again or rephrase the question. 'No' is sometimes easier for someone with Huntington's to say than 'yes' – it may mean 'not right now', 'I don't want to change what I'm doing' or 'I need a bit more time to process what's happening next'. It's important to make sure there is enough time in the care package for carers to work at the person's pace.
- **Training** - training about Huntington's, particularly on communication and the cognitive features of the disease can help carers support those with Huntington's. Bespoke training is available from the Huntington's Disease Association.

Expect repetitive insistence

The person with Huntington's disease may ask you the same question over and over. Even though you take the time to answer the question a few times, you may be asked again.

The person probably understands what you are saying, but has difficulty remembering, difficulty with anxiety or cannot change the topic they are focused on. Please be patient. Huntington's causes this behaviour. Try to gently change the topic.

Unable to see consequences

Perseverative thinking, in other words being “stuck” on an idea or thought, can make it difficult for someone with Huntington's disease to reflect on negative consequences of a decision they have made. They just “want what they want”.

People with Huntington's find imagining the future challenging and this may make capacious decision making difficult in relation to imagining consequences of their decisions.

They may act in an impulsive way, for example, spending money without considering the consequences or being generous to friends and family or even the “bloke down the pub”. Be aware that this impulsive behaviour makes them vulnerable.

Irritability and aggression

It is common for some people with Huntington's disease to be emotionally volatile. They may “flare up” for no apparent reason or over trivial issues. In this situation, it is best to avoid confrontation. If necessary, leave the room, particularly if there is the threat of physical aggression.

These episodes are often short lived and quickly forgotten by the person with Huntington's disease. Try and avoid subjects that may be triggers to this irritation or to overload them with too much information or too many questions at once.

Importance of routine

People with Huntington's disease may seem mentally inflexible. They like to adhere to set routines and behaviour patterns and appear unwilling to adapt to new situations or altered circumstances.

It is important that any care plan incorporates the need for as few carers to provide the care as possible. People with Huntington's struggle with change and facing new challenges. They are unable to instruct new carers to their needs daily and will quickly lose patience. This may cause conflict between them and their carer and lead to a breakdown of engagement and the care service provided.



Strategies to consider

- **A consistent approach and limited choice** - people with Huntington's need a very clear and set routine as the part of the brain that deals with choice and change is damaged by the disease. Due to the time it takes for people with Huntington's to process information and understand the consequences of any decisions made, it is helpful to keep information clear, concise and any choices offered limited.
- **Moving from one activity to another** - the transition from one activity to another (e.g. watching television to eating) can be difficult for someone with Huntington's. It is important to incorporate time to prepare the person about activities into the routine and make sure all involved follow this pattern.
- **Reinforcing the routine** - once a routine is established, the person with Huntington's may need to be prompted and reminded. This could be through regular verbal or phone reminders that something will be happening or visible visual charts that show the routine and what has been agreed.
- **Stay positive and don't push** - try to keep things positive, friendly and with a good sense of humour! Give time, and return to the issue later if needed. Return to the issue later if the person becomes frustrated or upset.
- **Keep a log** - it is a good idea to record any triggers or changes and share these with those involved with the individual to help maintain a consistent routine and approach.

Communication



As Huntington's progresses, the ability to communicate effectively decreases. People with Huntington's develop dysarthria, a difficulty with the production of speech and they find it difficult to think and process information. Giving time, being patient and communicating in a quieter environment are important.



Communication top tips

- Allow the person enough time to answer questions.
- Offer cues and prompts to get the person started.
- Be patient.
- Move the conversation to a quieter, calmer environment to avoid distraction.
- Ask the person to repeat phrases you don't understand or spell out the words.
- If the person is confused, speak more simply and use visual cues to demonstrate what you are saying.
- Consider using communication devices such as yes / no cards.
- Consider using technology such as a mobile phone or tablet.



Physical challenges

Big 'bursts' of movement

People with Huntington's are often affected by involuntary movements, known as chorea and the impairment of voluntary movements.

Although chorea is the most obvious symptom, it is usually the disturbance of voluntary movements that are more problematic leading to reduced dexterity, slurred speech, swallowing difficulties and problems with balance and falls.

People in the more advanced stages of Huntington's disease often have difficulty controlling their voice and movements. For example, when getting up from a chair the muscles in the legs may use more force than needed to lift them off the seat, giving the impression they are leaping out of the chair. Their voices may be raised, giving the impression they are angry.

Spatial awareness is also affected, and they may stand too close for comfort whilst talking to you. This can feel intimidating but is not intentional on their part.

Be aware of the risk of falls

By the time a person needs care in the home, it is most likely that their balance is impaired. People with Huntington's disease often walk about their homes holding onto furniture.

Assessment by an occupational therapist can help with the installation of rails and safety devices. It may also be helpful to be assessed by a physiotherapist at a local hospital. A GP can arrange this. The person may need to consider changing the layout of their home or even a change of housing to allow for further deterioration of their mobility.

See through “the disguise”

Weakness and changes in the tone of the facial muscles, often contribute to an appearance of boredom. Difficulties maintaining a smile while listening or speaking may make a person with Huntington's disease look unhappy, bored or uninterested.

Weakness and changes in posture, such as leaning to one side, may look like 'attitude'. Maybe you will think this person does not like you. Don't let this "Huntington's disguise" fool you! This person may be smiling on the inside, very interested in what you are saying to them and does like you. Don't give up!



Diet, swallowing and oral health

The importance of diet

Many people with Huntington's disease find it difficult to maintain their weight. Weight loss is a common feature of the condition and it is not yet fully understood why. It is in part due to metabolic changes. It may also be due to the involuntary movements using up extra calories, or it may be that swallowing problems mean the person avoids eating as it is so distressing for them. Low body weight leads to increased risk of infection, muscle wasting and lethargy.

A high calorie diet should be encouraged, and the advice of a dietitian sought. They may be able to recommend the GP prescribe supplementary high calorie drinks or give advice on how to increase calories in the diet.

Beware of the risk of choking

As the disease worsens, people with Huntington's develop a swallowing disorder. You may not be able to see that the person is having difficulty swallowing, but you need to be aware that they may need an adapted diet.

Some people with Huntington's disease eat too fast, forget to chew and over fill their mouths with food. This increases the risk of choking. They need plenty of time to eat their meal and may need assistance cutting food up or getting food to their mouths and monitoring whilst they are eating. Eating is more time consuming for someone with Huntington's disease and all this must be taken into consideration when planning care.

A Speech and Language Therapist should monitor the person and assess the ability to swallow regularly. They can make recommendations on adapting the diet.

There is a lot that goes into managing daily tasks. Not all of this can be seen. Let's take a closer look at eating and drinking....

Getting started

I need to remember to eat and drink, and my brain needs to get me started on this. I might need prompting to help me do this.

Monitoring risks around me

I need to stay safe. Is my food out of date? Can I remember what can safely go in a microwave? Is my food cooked properly? Is my food/drink too hot or cold? Am I able to keep an eye on food that is cooking in the oven or hob?

Managing more than one thing at once

It might be hard for me to cook a meal, particularly if I need to be doing more than one thing at once. If I get distracted while I'm preparing or eating my meal, I will find this hard and this will make it riskier for me

Preparing food

There are a lot of things I might need to do to prepare my meal - chopping, carrying, boiling, heating.

Memory

I need to remember everything I've been told which keeps me well and safe. Sometimes I might not be able to do this at the right time.

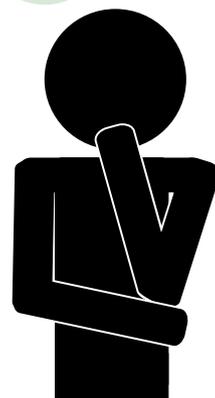
Planning and organising

I need to plan and organise my meals, and my shopping so that everything I need is available.

People with Huntington's have difficulties with all these things.

I need extra time

Preparing and eating my meals takes a long time. I might get tired and need to have smaller, more regular meals or come back to it later.



Importance of dental care

Involuntary movements make it difficult for someone with Huntington's disease to brush their teeth effectively. People with Huntington's often eat a very sugary diet, which may lead to problems with decay and to further problems with eating. If there is a swallowing problem, increased mouth bacteria from poor dental hygiene may increase the risk of chest infections and pneumonia.

As the disease progresses the ability to manage personal dental care can decrease, therefore it is very important that someone becomes responsible for this.

Smoking



If the person you are seeing with Huntington's disease still smokes cigarettes, you can assume that smoking is very important.

It is important to remember that they may have suffered loss after loss: their job, driving a car, friends, their place in the family and their ability to live independently. Smoking often takes on a symbolic importance as “the only thing I have left”.

Try and be tolerant in their home. You may ask them to refrain from smoking whilst you are in the house.

Remember, impulse control problems mean the person with Huntington's cannot wait...especially for a cigarette. They may forget your agreement regarding smoking in front of you.

People with Huntington's often cannot feel empathy and so appear to be selfish and only concerned with their own needs. Try to remember this is the effect of the illness and not personal to you.



Support and further information

Working with someone with Huntington's disease can seem quite daunting at first, there is so much to take in and understand. It will take a lot of kindness and patience to fully assess this person and you may have to rethink the ways you do things. The most important thing to remember is that they cannot adapt their behaviour to you, **you** must be the person who adapts to them. The person's family members can be an invaluable resource in the assessment process but may feel they cannot openly discuss the issues in front of the person with Huntington's for fear of upsetting them. Be tactful and flexible in your approach.

Specialist Huntington's Disease Advisers (SHDAs)

At the Huntington's Disease Association, we have a specialist advisory service operated by Specialist Huntington's Disease Advisers who have a background in health or social care and are knowledgeable about Huntington's disease. They operate throughout England and Wales and are able to provide bespoke training to staff working with clients with Huntington's disease. They are also able to discuss individual problems, provide tailored advice and suggest ways of managing certain behaviours. To get in touch with your local Huntington's Disease Adviser, contact us at:

📞 0151 331 5444

✉ info@hda.org.uk

Check the back page of this publication as the direct contact details of your local adviser may have been added.

Specialist Huntington's disease Youth Engagement Service (HDYES)

At the Huntington's Disease Association, we have a Youth Engagement Service (HDYES) operated by two youth workers who work with young people whose families are affected by Huntington's. This confidential service is for anyone aged 8-25 living in a family affected by the disease (including extended family such as a cousin, grandparent, etc.). If you are supporting a young person living in a family affected by Huntington's and you wish to refer them to HDYES, get in touch with your local Specialist Youth Worker at:

 0151 331 5444

 info@hda.org.uk

Website

Our website offers practical advice and sources of help and support, including downloadable information guides. We have guides with more in depth information about particular areas of Huntington's disease such as mental illness and mental capacity and care in advanced Huntington's. It also holds information about events and national training events that you can attend:

 www.hda.org.uk

Membership

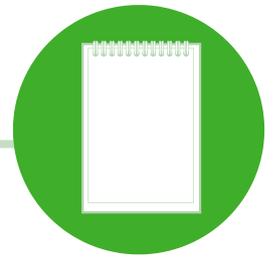
You can join the Huntington's Disease Association as a professional member and get access to expert information, early bird invitations to our events and our twice-yearly magazine / newsletter. After six months, you will become eligible to apply to become a voting member. There is a £30 lifetime membership fee to join. Further information about membership can be found on our website.



Resource library

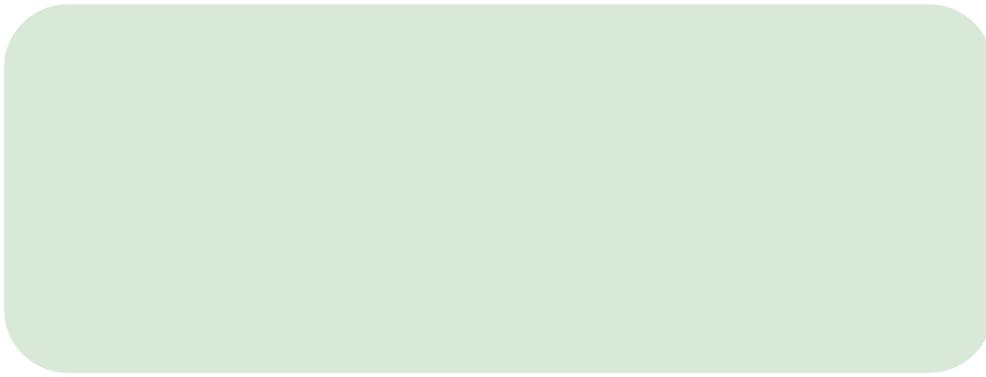
The following charities, organisations, publications and websites may be able to provide further information and guidance on the topics covered in this guide:

Organisation	Contact details	Support
Huntington's Disease Association	0151 331 5444 info@hda.org.uk www.hda.org.uk	<ul style="list-style-type: none">AdviceSupport resourcesOnline forum
HD Buzz	hdbuzz.net	<ul style="list-style-type: none">Research news
Huntington's Disease Youth Organization (HDYO)	info@hdyo.org en.hdyo.org	<ul style="list-style-type: none">AdviceSupport resourcesOnline forum



Notes

Local information



Get in touch

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

email **info@hda.org.uk**

phone **0151 331 5444**

www.hda.org.uk



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We'll be there