Huntington's Disease Association

# Cognitive changes in Huntington's disease

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# Contents:

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Dr Gunn has worked with the Huntington's Disease Association to develop and deliver new psychological therapies for people affected by Huntington's disease. Alongside leading a group of researchers into Huntington's disease based at Leicester, she promotes research into Huntington's as co-lead for the Leicestershire Huntington's Disease Network, which is a collaboration between families affected by Huntington's disease, charity partners, NHS services, and researchers.

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# Introduction

The cognitive symptoms of Huntington's disease can begin several years before the more obvious physical symptoms, such as involuntary, jerky movements (called 'chorea').

Cognitive changes become more severe over time and ultimately lead to sub-cortical dementia. These changes are likely to include changes in executive function (leading to disinhibited behaviour, risk-taking, loss of insight, and problems with planning and organisation), apathy, memory loss, attentional problems and disorientation.

These cognitive changes often have a substantial impact on daily functioning, which increases over time. It is essential to understand this group of symptoms, both in terms of being able to support someone with Huntington's effectively and to complete a full and accurate assessment of the person's needs.

Cognitive symptoms and their functional impacts can lead to emotional difficulties and behavioural changes. They can cause intense psychological discomfort, not only for the person with Huntington's, but for their caregivers.

This chapter gives an overview of the main cognitive changes that affect people with Huntington's disease and how they can be managed.

# Cognitive changes: An overview

The main cognitive changes experienced by someone with Huntington's disease are:

- **Changes in executive function:** Executive function serves as the brain's management system, overseeing other aspects of cognitive function. Executive function includes skills around planning and organisation, emotional regulation and cognitive flexibility (which includes adapting thinking and behaviour to changing situations, such as switching attention between tasks and managing competing demands). Perseveration is a common difficulty in which a person with Huntington's can "get stuck" on a particular thought or action, and struggle to disengage from it.
- **Changes in social cognition:** People may struggle with social cognition, leading to challenges in interpreting and conveying emotions. For example, people with Huntington's may struggle to gauge a person's mood based on their facial expression. These difficulties can cause increased vulnerability, as people may be less able to assess risks in their environment. The changes can also impact personal relationships.
- **Development of apathy:** People with Huntington's often become less likely to initiate goal-directed behaviour and may stop engaging in previously valued activities or relationships. Apathy in Huntington's appears to be multidimensional, and people may experience some of these difficulties but not others.
- **Changes in memory:** Memory challenges often lead to difficulties in acquiring new information or recalling previously learned material. In the later stages of Huntington's, people may also become disorientated and might be confused as to where they are, the date, and / or who the people around them are.

- **Bradyphrenia:** People may experience slowing of their thinking processes and response times, which can create risks and impact on communication.

Cognitive changes have extensive potential impacts, and may lead to the person:

- Avoiding social interactions and activities that they would usually enjoy, which can lead to loneliness and isolation
- Not engaging with health and social care workers
- Encountering conflict in the family or with friends
- Encountering challenges at work
- Being more vulnerable to exploitation (e.g., in relation to finances)
- Being at risk of stigmatisation and damaged relationships, if deficits in social cognition result in inappropriate responses to social cues
- Encountering a wide range of functional difficulties with daily tasks
- Self-neglect and having poor hygiene due to a lack of awareness or initiation around self-care
- Taking actions which are unsafe or unwise, because the person does not understand the extent of their difficulties (or may think that they are not experiencing difficulties at all)

Lack of insight (anosognosia) regarding symptoms, including cognitive changes, is common among people with Huntington's. This can make it especially challenging for health and social care professionals to assess the person's needs accurately and provide the appropriate care. The Huntington's Disease Association has a guide (see 'resources' below) for both families and professionals on important things to be aware of when working with someone with Huntington's, as well as practical tips on how to offer support.

# Assessing cognitive changes in people with Huntington's disease

Cognitive changes caused by Huntington's disease affect people differently. It is important to take the time to understand a person's specific difficulties and how they are impacting their life and wellbeing. This can vary significantly between different individuals.

Assessing cognitive changes can help to better understand a person's needs and how to meet them effectively. The table below (Stoker et al, 2022)

provides an overview of some cognitive and neuropsychiatric measures, which can be used to assess cognition. This is not an exclusive list, but provides some initial options to consider.

**Table 4 - Cognitive and neuropsychiatric measures useful in the assessment of Huntington's disease**

| Domain | Tests |
|---|---|
| Global Cognition | • Addenbrooke's Cognitive Examination–III<br>• Montreal Cognitive Assessment |
| Executive function | • Symbol Digit Modalities Task<br>• Stroop test<br>• CANTAB–One Touch Stockings of Cambridge |
| Learning and memory | • Hopkins Verbal Learning Task<br>• CANTAB–Paired Associates Learning |
| Attention | • Wisconsin Card Sorting Test<br>• CANTAB–IDED |
| Language | • Phonemic verbal fluency<br>• Semantic verbal fluency |
| Depression | • Hospital Anxiety and Depression Scale<br>• Beck Depression Inventory |

| | |
|---|---|
| Apathy | • Apathy Evaluation Scale |
| Social cognition | • Toronto Alexithymia Scale<br>• Empathy Quotient<br>• Reading the Mind in the Eyes |



**Source:** Stoker, T. B., Mason, S. L., Greenland, J.C., Holden, S.T, Santini, H., Barker, R. A. Huntington's disease: diagnosis and management. 2022. BMJ Journals - Practical Neurology.

# Changes to speech, language and communication

The cognitive impacts of Huntington's disease can affect speech, language and communication in the following ways:

- **Speech:** A person's speech can become slowed / slurred (signs of 'dysarthria'), with inappropriate pauses or bursts of speech.
- **Language:** Verbal fluency can lessen, with people struggling to generate words (spoken or written / typed). It can become harder to understand and produce complex sentences. This may be particularly pronounced when there are competing cognitive demands (e.g. in noisy environments or when multiple people are speaking at once). These symptoms are usually most severe during the later stages of Huntington's.
- **Communication:** Communication changes are impacted by the cognitive changes described above, including difficulties with organising thoughts and recalling the information needed for the current conversation. Slower speed of processing also impacts communication; people with Huntington's generally need longer to process information provided to them and to prepare a response.

# Clinical management of cognitive symptoms

There is a range of approaches to managing cognitive symptoms. It is crucial to tailor support to the person's needs, rather than taking a prescriptive approach. Some possible approaches include:

- Multi-disciplinary rehabilitation strategies (speech therapy, occupational therapy, neuropsychological or clinical psychological input) might support the person to cope more effectively with their cognitive difficulties. Co-working between disciplines in joint or parallel sessions can be helpful; for example, collaborative work between speech therapy and psychology can help if a person is feeling anxious or self-conscious about changes to their speech
- Using strategies to manage behaviour may help with maintaining positive relationships with family and friends. These behavioural strategies may be most effective when co-developed with the person with Huntington's and their close family or healthcare workers who support them. It may be that the person with Huntington's is able to put strategies in place for themselves (possibly with support), or their caregivers might take the lead
- Relationships (with the family, professionals and others) can be helped if those supporting the person with Huntington's understand the condition, understand why someone may be behaving differently, and know or develop strategies to support the person
- Support from friends, family and services can help to retain social opportunities or develop new ones
- Treatment for anxiety and depression may help to mitigate the impacts of some types of cognitive change
- Cognitive stimulation and co-developed strategies through rehabilitation may improve or help to maintain planning and initiation over the short term
- Sedative drugs and neuroleptics should be chosen with caution, and their effects closely monitored, as they impair executive function and attention
- Patient / family education and structuring the person's home environment can help to mediate the effects of cognitive changes. The

person with Huntington's can be supported to add tools and strategies into their daily life that help them rely less on their internal cues or prompts from others, thereby helping to maintain an appropriate level of independence. This can be particularly important with essential health issues, such as the person forgetting to eat regularly. Often, these are things that we all use, but which become more important if someone is experiencing cognitive difficulties. Examples include:

- Setting reminders on a phone (if the person is able to use a phone, or can be helped to do so)
- Keeping a small whiteboard on the fridge with 'things to remember today'
- Setting up recurring payments for household bills if someone is struggling to manage their finances

These can be quite small changes, which may make a significant difference once established in regular use.

# Personalised care planning

Cognitive changes can be managed more effectively by:

**Asking questions in a helpful and accessible way:**

The impact of cognitive changes means that people with Huntington's disease can struggle with making complex choices. Ask specific and realistic questions. For example, rather than asking 'What do you want to do about keeping the house clean?', try 'We would like to try a cleaner for your house, do you think that's a good idea or a bad idea?'. This allows the person to think about a relatively simple initial response (and also avoids leading them to a particular answer to support their autonomy). Follow-up questions can be used to encourage the person to elaborate.

**Understanding why a person is not engaging:**

Over time, Huntington's disease makes it harder to plan, organise, initiate activities, and problem solve. People may not always respond to calls, letters or visits. This should not be taken as a refusal to engage – it may be a sign of disease progression. If you have the opportunity, take time with the person to explore the reasons why they are not engaging. Consider the following questions:

- Are they struggling to access appointments?
- Do they need practical help to attend appointments that they are not getting?

- Is travelling to appointments financially difficult?
- Is someone preventing them from attending?
- For home visits, are they embarrassed to answer their door because the house is not tidy or clean, or because of issues relating to personal hygiene?
- Do they worry their difficulties might not be understood, or that they may be judged or criticised?
- Are they feeling hopeless and questioning the point of attending appointments?

Consult with other members of the multi-disciplinary team and ask the person appropriate questions about their lack of engagement, if possible. It may be that the person truly does not want to engage with the service - if they have the capacity to make that choice, then that is up to them. However, try every route with patience before coming to that conclusion.

**Being willing to adapt communication methods:**

Consider contacting the person at different times of the day and using different methods. Some people prefer text or email communication, because it gives them time to process information and respond at their own pace. However, from a safeguarding perspective, recognise that it could be someone else sending / responding to texts and emails. Always make an agreement with the person first that it is acceptable to communicate with them in this way. Other people may prefer to speak directly. When doing so, give them lots of time to think and respond. What feels like a very long pause may be crucial thinking time for the person with Huntington's, which enables them to have a conversation. Remember that every time you speak, you are 'restarting the clock' as the person processes what you have said. Therefore, it is better to keep quiet while the person thinks it over (resisting the common temptation to try and rephrase / simplify what we have said, to help the person understand).

**Planning home visits effectively:**

Home visits are important to help build a relationship and understand a person's living situation. They provide an opportunity to observe and respond to the person's body language, and for them to do the same with yours. It also allows you to be confident of which other people are present (not always possible on a phone call or video call). Persistence is often required, as people may find it difficult to keep track of appointments. Patience is also crucial to gain trust.

People with Huntington's disease can find it difficult to deal with changes of

plan or unexpected events, such as postponing a meeting or changing who will be attending. Giving adequate notice of a visit and not changing the agreed time or arriving late can help to build trust. Consider whether an initial joint visit with a person they trust would be beneficial to building your relationship with them. Remember that people with Huntington's may have experienced stigma or a lack of understanding from healthcare staff and other professionals in the past, meaning it can take time to earn their confidence.

**Understanding what makes a person irritable, frustrated or angry:**

Some people with Huntington's disease may become irritable, frustrated or angry in response to certain triggers (these may not always be obvious). They may struggle to contain these feelings at times. This can be challenging for family members, as well as for professionals and care providers working with the person. Consider taking a colleague with you to assessments for support, where an increase in agitation or volatility has been highlighted. Check in with caregivers regularly about how they are coping to ensure everyone's safety.

Frustrated or angry behaviour is generally rooted in understandable causes. Take time with the person to understand their experiences. The person may be feeling stressed, angry or in grief about their condition and the limitations it can bring; they might also be hungry, in pain, or otherwise uncomfortable (e.g., too hot or too cold). Understanding and avoiding a person's triggers for irritable or aggressive behaviour is key to good communication and the acceptance of support. It may also support the person to self-regulate if you develop a formulation of their triggers together. For example, it is common to find it frustrating if offered a choice that then becomes unavailable. In the context of Huntington's, one example might be offering a specific care home, which then turns out to be too expensive. Cognitive changes linked to Huntington's can exacerbate this understandable irritation – a person may become preoccupied with one option and struggle to accept any alternative, which can then be a trigger for anger if that option becomes unavailable. Accordingly, options should not be offered without a good degree of certainty that they will be accessible.

**Being aware of cognitive changes when carrying out an assessment:**

Assessing a person with Huntington's disease can take time due to communication difficulties and slowed processing of information. Do not necessarily take their first response to questions at face value (especially if it is very brief, such as a simple 'yes' or 'no' to a complex question). If you are unsure, check with them that they have understood. Avoid covering

multiple issues in one short assessment, as too much information in a brief period can be overwhelming for someone with Huntington's. Limit it to one or two topics, pace the delivery of information, and check regularly that the person is following and understanding you. It may be helpful to use visual aids to support the person's understanding.

Remember that people may take longer to complete tasks due to slower processing and will likely need extra time. This is especially important to consider when arranging a care package. Care packages risk failing if they do not provide adequate time for a person with Huntington's disease to complete tasks. Be aware that people with Huntington's may not recognise the extent of their difficulties, or even acknowledge them at all (the anosognosia; described above), so seek confirmatory information from caregivers if in doubt.

**Knowing how to respond when someone becomes fixated on something:**

Sometimes people with Huntington's can get 'stuck' on certain ideas and find it difficult to move on. This is known as perseveration. For example, a person could perseverate regarding a particular item, such as bottles of water, and buy them in excess. They may also ask questions on the same subject repeatedly, which can be a source of anxiety for the person and stressful for those supporting them. Consider talking about this difficulty if the person is likely to be receptive to co-developing some strategies to manage perseveration. If this is not possible, then working with caregivers to plan responses that minimise anxiety but reduce their load may be helpful.

Someone with Huntington's disease may have compulsions that take the form of repetitive behaviours and actions that are associated with an obsessive thought. They may also experience intrusive thoughts that cause them anxiety. Arguing with the person with Huntington's or trying to stop them from carrying out these repetitive actions is unlikely to be effective; they probably will not understand why you think they are being unreasonable, and it will likely damage the relationship between you if you try. Distraction can sometimes be helpful, but it is important to be realistic and (provided the person is not at risk) it can be best not to intervene.

# Conclusion

Cognitive symptoms for people with Huntington's can be challenging to manage for the person, their family, and the health and social care professionals supporting them. However, there are ways to manage symptoms that can reduce anxiety and stress for all involved – most importantly, the person with Huntington's – and to ensure that an assessment reflects the person's needs accurately to help get them the right support. Most crucial is a good understanding of the importance of the cognitive changes which come with Huntington's and maintaining a person-centred approach to understanding and supporting each person.

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# Resources

### Huntington's Disease Association resources

### For professionals

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

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

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

### Stay informed

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

### For people affected by Huntington's disease

Our information resources are available at **https://www.hda.org.uk/information-and-support/information-resources/**

### Specific resources relating to cognitive changes are

Behaviour and communication guide. Available here.
*N.B. This is a useful resource for families, which covers some strategies for cognitive changes.*

Dealing with lack of awareness. Available here.

Hurry up and Wait: A cognitive Care Companion – Huntington's Disease in the Middle and more advanced years. Jimmy Pollard. Available here.
*N.B. Pollard has written this for carers, to help manage the day-to-day challenges caused by cognitive changes.*

### Support for children and young people

Find information on our Huntington's Disease Youth Engagement Service (HDYES) by searching for "HDYES" on the website.

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## Get in touch

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

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