Huntington's Disease Association

Mental health treatment and support for adults who have Huntington's disease

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Introduction

Huntington's disease is an inherited neurodegenerative condition which causes progressively severe difficulties with movement and thinking (cognition) and can be associated with worsening mental health.

People living with Huntington's disease will often need support from mental health services. Although brain changes may be associated with declining mental health among people with the disease, a holistic formulation of any mental health difficulties is essential.

These guidelines aim to offer healthcare professionals working in mental health services an overview of mental health symptoms and difficulties experienced by adults with Huntington's disease, and guidance on how to offer effective support.

People with Huntington's disease commonly experience anxiety and low mood (which may be diagnosed as depression). They can also develop changes in their personality, which can contribute to irritability, challenging behaviour, disinhibition and apathy. Rarely, patients with Huntington's disease may experience psychosis. People with Huntington's disease, and their family members, often report mental health symptoms and difficulties as being as important as its impacts on physical health.

Many people with Huntington's disease may develop a degree of lack of awareness or insight into their condition ('anosognosia'). This can include not being aware of how the disease has affected them, their mental health and the people around them.

Many of these symptoms and signs of distress can respond well to intervention, including psychological and medication-based treatment options. However, despite the importance of mental health support in Huntington's disease, patients are often rejected from local services. This means that mental health symptoms in Huntington's disease are at risk of under-diagnosis and under-treatment.

People with Huntington's disease have a right to be seen and treated by local mental health services. Although the underlying condition is not yet curable, mental health support and treatment can have a tremendous impact on the quality of life of patients and carers. These guidelines aim to offer a starting point for providing this much-needed care.

General principles of mental health support for adults who have Huntington's disease

It is crucial that any professional treating a person with Huntington's disease has an understanding of the condition and its potential impact on mental health. While Huntington's disease has several unique clinical features which must be considered, the approach taken towards the assessment and treatment of people with other mental health diagnoses is still highly relevant and helpful. Many mental health presentations in Huntington's disease can often be treated using similar core principles routinely used in mental health services.

Stage of Huntington's disease and relevance for mental health

People carry the Huntington's disease gene expansion from pre-birth and can choose to undergo genetic testing to confirm the genetic diagnosis, meaning that the person carries the gene expansion and will develop the disease later in life (usually aged between 30 to 50 years). Testing is generally not available until a person is an adult (aged 18 years or older). Huntington's disease is clinically diagnosed when clear movement symptoms are evident on clinical examination. Prior to the development of these movement symptoms, people who carry the faulty Huntington's disease gene are considered 'pre-symptomatic'. The transition from pre-symptomatic to symptomatic Huntington's disease is slow and gradual.

Avoid 'diagnostic overshadowing' – the assumption that carrying the gene expansion for Huntington's disease is the biological cause of any mental health difficulty, even before other features develop.

The emergence of some of the features of Huntington's disease may contribute to mental health difficulties in pre-symptomatic people carrying the Huntington's disease gene expansion (especially if they are close to predicted onset).

Be aware that grief, distress, worry, and social and functional losses associated with Huntington's disease can understandably contribute to worsening mental health. For example, taking early retirement or losing a job due to the disease can cause significant distress. Due to its inherited nature, people may have experienced multiple bereavements and be acutely aware of how their disease will progress. It is also common for the person and their family members to grieve for anticipated future losses, such as not having the career they wanted or having to give up enjoyed activities.

Societal pressures and stigma can contribute to poorer mental wellbeing. Many people will have experienced a parent (and possibly other family

members) having Huntington's disease, and they may have cared for them too. Some people will have grown up knowing that they had a 50% chance of inheriting the disease; others may have learned about its presence in their family more recently.

The focus of this guideline is on people who are living with *clinical* Huntington's disease. However, there may be some overlap with pre-symptomatic people carrying the Huntington's disease gene expansion, particularly for those who are close to developing symptoms / signs of the disease.

Types of mental health conditions commonly seen in people with Huntington's disease

Common mental health conditions and risks in Huntington's disease

People with Huntington's disease are at increased risk of being diagnosed with a variety of mental health conditions.

Common mental health conditions which may be diagnosed in people with Huntington's disease include depression, generalised anxiety disorder, panic disorders and phobias. Substance misuse may also be identified. Transdiagnostic difficulties such as loss of identity, shame and low self-esteem are often present and can sometimes be overlooked. People living with Huntington's disease may be reluctant to leave their home if they are concerned that others would notice their movement difficulties or be negatively affected by their cognitive or personality changes. Be aware that the person may also feel overwhelmed in busy or noisy environments, leading to an avoidance of social situations. This can create a cycle of decreasing communication and social isolation.

Be aware that some aspects of Huntington's disease can resemble clinical features of mental health conditions. For example, 'apathy' (i.e., apparent loss of motivation and reduced goal-directed behaviour; more on this below) may present in a way reminiscent of depression, but without associated low mood or loss of enjoyment. Increased cognitive rigidity and perseveration can resemble obsessive-compulsive disorder (OCD), but lack the distressing / anxiety-provoking element, which is common in OCD. An example of perseveration is when a person gets 'stuck' on certain ideas and finds it difficult to move on. For instance, they might ask questions on the same subject repeatedly, which can be a source of anxiety for the person and

stressful for caregivers.

Identifying psychological difficulties and psychiatric symptoms correctly is a key skill that mental health services can provide to support people living with Huntington's disease.

Low mood (depression)

It is common for people with Huntington's disease to develop low mood, and some may reach the threshold to be formally diagnosed with depression. It can be difficult to distinguish between depression and the impact of other difficulties common to Huntington's disease. These include sleep disturbance, apathy, and physical slowing of movements.

While low mood seems at least partially associated with neurological changes, it is important to consider wider personal and systemic causes. People may understandably feel distress and sadness associated with the progression of their symptoms, disruption to relationships, and loss of independence and valued activities.

Anxiety

People with Huntington's disease commonly experience anxiety, which may arise in multiple forms, including panic and generalised anxiety, as well as specific fears. This can cause substantial distress and be difficult for family members, as such anxiety can become intense and overwhelming.

Worry about the future is understandably common, including fears of future loss of function, cognition and sense of self, and loss of independence. The person may also have potential financial and housing concerns, and fear stigma and discrimination. The impact of the disease on family members can be a significant source of worry. This can include a fear of becoming a burden as the person's needs increase, as well as genetic concerns related to the risk of children inheriting the disease.

Anxiety is an understandable difficulty to experience due to the significant stressors and losses the person will face. Past experiences of Huntington's disease in the family can contribute to anxieties around the future and engaging with health services. Many people may have seen older family members struggle with the condition in the past and worry that these difficulties will be replicated for them in future.

Psychosis

Psychosis is relatively rare in Huntington's disease, but some people do experience it. This can appear as delusions and hallucinations, similar to those associated with diagnoses of psychotic conditions such as schizophrenia.

Psychosis in Huntington's disease can occur in association with severe depression or other mood disorders. Huntington's disease is a slowly

progressing condition and sudden changes are very unusual. Consider the possibility of delirium in cases of new onset psychosis in Huntington's disease; delirium or acute confusion can be caused by infection (such as sepsis or a urinary tract infection (UTI)), constipation or a recent fall.

Psychosis in all conditions, including Huntington's disease, is often associated with a loss of awareness or insight. This can shape the risk assessment and need for treatment, as the person may not wish to cooperate with any plan made, or not realise that they are experiencing psychosis at all.

Organic personality disorder

People with Huntington's disease may experience changes in their personality, secondary to the condition and to the stresses and difficulties associated with it. Typically, this personality change can lead to increased irritability and behaviour which appears aggressive (verbally, physically, or both). Also associated with personality change seen in Huntington's disease is apathy. This can lead to people with Huntington's disease not initiating or responding to actions, seeming flat or blunted in their emotions, or appearing socially and emotionally disengaged. Apathy and irritability are common and may occur together.

'Organic personality disorder' is a type of personality change caused by physical damage or dysfunction in the brain, particularly in the frontal or temporal lobes. People with organic personality disorder may be impulsive and inflexible. For example, a person might make an impulsive, unconsidered purchase of a car that they cannot afford; they may also make socially inappropriate remarks (e.g., making personal comments about a person who is within earshot). The person may lack insight into these alterations in themselves, which makes it more difficult for them to manage their own behaviour or for others to support them with this.

In Huntington's disease, a diagnosis of organic or secondary personality disorder represents a change in previous personality characteristics and seems to be related to a progression in the underlying condition.

As with other 'non-organic' personality disorders, these changes are persistent, often chronic, and not explained by other mental or physical health problems. They can cause significant interpersonal difficulties and impairments in functioning and may be associated with risk to self and others.

In rare cases, where a person with organic personality disorder is refusing treatment but there are very significant risks (such as violence to self, harm to others or marked self-neglect), it may be necessary to consider treatment under the Mental Health Act (1983). In these situations, be aware that even though the personality change is secondary to Huntington's disease, it is still considered a mental disorder for the purposes of the Mental Health Act.

Dementia in Huntington's disease

This diagnosis is usually reserved for people with Huntington's disease who have problems with cognition over several areas, including memory. It refers to a global progressive impairment of cognitive function affecting functioning and is a common feature of advanced Huntington's disease.

Assessing the mental health needs of a person with Huntington's disease

The approach to assessing the mental health needs of a person with Huntington's disease is broadly similar to that of other patients, but with important additional considerations. Consider the following before and during the assessment process:

The relationship between cognition, movement and mental health difficulties

Cognition, movement and mental health difficulties often interact in somebody with Huntington's disease. Changes in behaviour may not be indicative of a mental health condition. For example, frustration related to an inability to communicate or to requiring support with activities previously done independently may lead to irritability and anger. However, this would not be due to a personality change or a diagnosable condition, rather to understandable frustration at new limitations.

The impact of trauma

Many people will have seen one of their parents live with Huntington's disease. Be aware that they may have experienced traumatic events in their history.

Taking a multi-disciplinary team approach

The care of people with Huntington's disease requires different specialties. When undertaking an assessment, involve local neurology services (or

specialist Huntington services if available), the Huntington's Disease Association and local community neurorehabilitation services.

Support for caregivers

Consider signposting / offering support to caregivers as part of the mental health support offered to the person living with Huntington's disease. Be mindful that family members are likely to also be coping with difficult emotions and stressors, which can include caring for several generations of people with the disease.

Offering support based on clinical need

There is no basis to exclude patients from mental health services just because of a diagnosis of Huntington's disease, especially in the context of significant mental health needs. Although mental health teams may not be specialists in Huntington's disease, they have specialist skills that can play an important role in improving quality of life. Seek advice from Huntington's disease specialists where needed.

Managing risks related to suicide

The risk of a person deliberately ending their life can be significantly elevated in people with Huntington's disease, highlighting the need for ongoing assessment and treatment of depression, anxiety, and impulsivity. A person's thoughts around ending their life may be in response to fears about loss of control and the future progression of their condition. Be aware that the person may have experienced the death of family members from Huntington's disease, which can cause worry about their own future. Take an empathic and non-judgmental approach to understanding and discussing any thoughts of this type. Recognise that a person with Huntington's disease is facing a deterioration in independence, wellbeing and self that is likely to be frightening and distressing. Should the person raise these issues, take the time to explore them empathically. Be mindful that it may not be inherently unreasonable for a person to have suicidal thoughts, and that thinking about it is not the same as planning to act on it. Use safeguarding approaches as appropriate to ensure the person's wellbeing and manage risk.

Being aware of risks associated with medication to treat movement symptoms

Medications used to manage movement symptoms in Huntington's disease may contribute to worsening mental health (more on this below).

Pharmacological treatment of mental illness in Huntington's disease

There are currently few evidence-based recommendations or randomised controlled trials to guide the treatment of mental illness in Huntington's disease.

People with Huntington's disease who have significant mental health needs will typically respond to the same pharmacological treatments as other people who have similar presentations. There are expert guidelines available, including in the Maudsley Prescribing Guidelines in Psychiatry (see References section below).

Be aware of possible side effects, which may worsen movement problems, stability or cognition. Atypical antipsychotics, especially at higher doses, might be associated with poorer cognitive function and physical health side effects, such as low blood pressure.

Ensure that excessive medication use, or polypharmacy is prevented. As with any patient, the aim should be to find the minimal effective dose.

The treatment of somebody with Huntington's disease in the context of core psychiatric presentations, such as depression, anxiety and psychosis, should follow NICE guidelines for each of these diagnoses while considering the points above.

Antidepressants such as SSRIs (Selective Serotonin Reuptake Inhibitors) / SNRIs (Serotonin-Norepinephrine Reuptake Inhibitors) and atypical antipsychotic medications (such as olanzapine and risperidone) can usually be safely used in Huntington's disease to manage mental illness.

The management of movement symptoms in Huntington's disease is not within the expected remit of a local mental health service. However, it is important to be aware of the following:

- Tetrabenazine, a common neurological medication for the treatment of excessive movements, is associated with a range of side effects, including low mood and suicidality. Consider stopping tetrabenazine for somebody experiencing depression who is prescribed this medication; make a decision in consultation with the local neurology team or Huntington's disease specialists.

- Atypical antipsychotics, such as olanzapine and risperidone, are commonly used to manage chorea and excessive movement (however, they are not licensed for this purpose). These medications may be advantageous in some cases if multiple symptoms can be managed by a single medication. Be aware, therefore, that if antipsychotic medication is stopped for mental health reasons, this may affect the control of movement symptoms.

Treating personality changes

Consider non-pharmacological strategies when managing personality change. Educating caregivers about the personality changes seen in Huntington's disease can help them change the environment, which, in turn, might mitigate difficulties.

Reducing demands on the person and keeping to regular routines can often help to reduce irritability.

A number of issues can contribute to irritability in people with Huntington's disease, including:

- Frustration with communication difficulties
- Changing abilities and reduced independence
- Fatigue
- Pain
- Hunger
- Constipation
- Infection
- Tiredness
- Feeling too hot or too cold
- Busy, chaotic environments
- Cognitive overload, often caused by delays in processing information leading to difficulties engaging in conversations

These causes will lead to different approaches to intervention, which may include resolving the underlying issue when possible, or at least working to minimise distress if not.

If other underlying causes of irritability have been ruled out and psychosocial interventions have been used, consider using SSRIs to treat irritability. If ineffective, especially if there is aggressive behaviour, mood stabilisers and atypical antipsychotics can be used to reduce irritability.

Excessive preoccupation with single issues, thoughts or behaviours (perseveration) can sometimes also respond to SSRIs.

Admission to mental health units

The severity of mental health symptoms and difficulties experienced by people with Huntington's disease may sometimes require admission to a mental health unit. This may particularly be required for people with significant psychosis, aggressive behaviour, or high levels of risk to themselves. Such admissions can be very challenging and stressful for all involved, but can also be both necessary and lifesaving.

While there are specialist inpatient mental health services for people with Huntington's disease, these are rare and often require specialist commissioning, and so cannot be accessed in an emergency. If the person is admitted to a mental health ward, ensure the following steps are taken:

- Early reviews by clinical psychologists, social workers, ward physiotherapists, occupational therapists and speech and language therapists where available (alongside the psychiatrist support which should be provided).
- Referral to the Huntington's Disease Association for support.
- Contact made with the person's specialist Huntington's disease service or local neurology team for additional support and guidance.

Psychological interventions for mental illness in Huntington's disease

There is a lack of research into psychological interventions for people with Huntington's disease. Adopt a holistic approach, informed by the person's specific difficulties and context, such as the people in their life, their home, and their networks.

People with Huntington's disease should not be excluded from psychological therapies. Evidence suggests that therapeutic options can be valued and seen as potentially useful treatment options by people living with Huntington's disease.

Be aware that cognitive changes may affect the person's ability to engage in therapy (see the Huntington's Disease Association Cognitive guidelines here). These can include difficulties in executive function (planning, impulse

control, cognitive flexibility), attention, memory and social cognition. Take an inclusive approach by trying to support engagement as fully as possible.

A limited number of therapeutic options exist for people with Huntington's disease, with some early evidence of effectiveness for some. This includes:

- In-person mindfulness based cognitive therapy
- Online group therapy programmes based on Acceptance and Commitment Therapy (ACT)
- Self-guided therapy for anxiety

Be aware that not all of these therapies are currently available, and some may have limited availability. Some were also designed for and trialled with people who are 'pre-symptomatic' (before physical symptoms of Huntington's disease begin). Further, not all modalities will be helpful for all people with Huntington's disease. Some people may not wish to (or be able to) access group therapy due to physical or cognitive difficulties; anticipating stigma or other anxieties may also prevent engagement. Self-guided therapy may not be accessible or helpful for those with high levels of apathy or memory difficulties. In all cases, more evidence is required regarding effectiveness, and the evidence is at an early stage. However, initial findings are encouraging and suggest that psychological interventions have a significant potential role to play in improving wellbeing for people living with Huntington's disease.

Given the low level of current evidence, refer to general guidance published by NICE for more common difficulties, such as depression in individuals with a chronic physical health problem. Refer also to the British Psychological Society (BPS) guidance on psychological interventions for people with Huntington's disease (see References section below).

For irritability, consider formulation-based approaches using therapies generally recommended for anger. This could include cognitive-behavioural or mindfulness-based approaches.

Broader care of a person with Huntington's disease

Multi-disciplinary team care

Mental health services play an important role in the care of people living with Huntington's. However, care should also be provided by a wider team.

Working collaboratively with other professionals is essential to meeting the needs of someone with Huntington's disease. Their needs may fall into several areas and will change over time.

Many people struggle to keep track of different professionals' involvement, especially when they are receiving support from multiple health and social care services. Avoid relying on the person being able to accurately recall who is involved with their care and what input they are receiving, not least because (depending on their stage of Huntington's disease) they may be experiencing difficulties with recall.

There must be robust communication and coordination among healthcare professionals, social workers, and other relevant services to provide comprehensive support. Alongside mental health support, people with Huntington's disease may receive support from:

- Speech and language therapy for assessments of speech and swallowing, including the risk of choking
- Dietetics for maintaining weight, and if percutaneous endoscopic gastrostomy (PEG) feeding is being considered
- Physiotherapy for, among other things, falls prevention and balance
- Occupational therapy for assessment of abilities around functional tasks
- Social services for advice and help on social and care issues, including funding for supported housing, care packages and respite services
- Local neurology services for review of neurological symptoms and chorea management, if no Huntington's disease service exists locally
- Palliative care services for end-of-life issues and symptom management
- Prison, forensic and probation services (where appropriate and required)
- Huntington's Disease Association Specialist Advisers

Regional specialist Huntington's disease clinics – typically with neurology and neuropsychiatry input – often cover large geographical areas and are based in a hospital setting rather than the community. While many patients may be managed in such services, these may not be able to provide local or outreach input.

Consider coordinating a multidisciplinary team meeting to ensure that the relevant staff are involved and working together. Ensure that caregivers have access to their own assessments and support, as they may also experience significant mental wellbeing difficulties.

Ensure that the most appropriate team member is involved, if there are wellbeing concerns (e.g., the person missing appointments, a change in the

household, or caregivers becoming ill). Non-attendance and apparent disengagement should not lead to discharge from mental health services - they should be recognised as part of the challenges associated with Huntington's disease. Be aware that there can be difficulties in getting a person to an appointment, which may include financial and practical barriers, and caregiver (un)availability. Do not assume that a lack of attendance automatically means a wish not to engage.

Adult safeguarding

There may be additional factors to consider around adult safeguarding when working with people with Huntington's disease.

Due to cognitive changes, a person may display more risky behaviours or be at higher risk of drug or alcohol misuse than the general population. Difficulties with emotional regulation, problem-solving and impulse control may cause conflict in personal relationships, sometimes leading to issues with antisocial behaviour.

Cognitive changes can also make it harder to manage finances, leading to increased risks around budgeting and debt, and vulnerability to coercion and financial exploitation. A person's lack of insight into their symptoms can lead to problems with seeking or accepting essential support (discussed below). This can lead to self-neglect, impacting personal health and hygiene, and the home environment.

Empowerment and advocacy

Advocate for the rights and autonomy of people with Huntington's disease, involving them in decision-making processes as much as possible and respecting their changing mental capacity. Help them to retain as much autonomy as possible – even if it is on seemingly insignificant decisions.

Consider the impact of cognitive changes on a person's ability to engage in decision-making processes, so that appropriate support can be provided. Try to engage the person in choices to the best of their ability. Language support tools or technology may be helpful if the person is struggling to speak or write – this may include 'Alternative and Augmentative Communication' (AAC), for which speech and language therapy support is crucial.

Encouraging discussions around advance planning can help people to make their own decisions for the future while they have the capacity to do so, such

as by making a Lasting Power of Attorney (LPA) or an advance decision to refuse treatment (also known as a 'living will').

Personalised Care Planning

Supporting decision-making

Many people with Huntington's disease struggle with making complex choices and can find it hard to plan, organise, initiate activities, and problem-solve. There are a range of ways to support people to engage in decision-making, which may be helpful.

As described above, people may not always respond to calls, letters or visits. This should not be taken as a refusal to engage. 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. Home visits are important to help build a relationship and understand a person's living situation. Persistence and empathy are often required, as people may find it difficult to keep track of appointments.

People with Huntington's disease can find it difficult to deal with changes or unexpected events, such as unannounced visits or a change of plan. Give adequate notice of a visit and avoid changing the agreed-upon time. This can help build trust.

Being aware of a person's triggers is important, as this affects the likelihood and success of involving people in potentially difficult conversations. Some people with Huntington's disease may become irritable, frustrated or angry in response to certain triggers (such as becoming aware of new limitations or losses, or discussing their symptoms). They may struggle to contain these feelings at times. This can be challenging for family members as well as for professionals. Try to understand and avoid these triggers to promote good communication and the acceptance of support.

Presenting information

When presenting information to consider for a decision, consider providing a written list of available options. People with Huntington's disease may struggle to independently generate suggestions, while still being able to choose between presented options.

Do not offer choices (e.g., a specific care home) if it is not certain they will be available. A person with Huntington's is at risk of becoming attached to an option, then struggling to accept any alternative if it's ultimately unavailable.

Conducting assessments

Assessing a person with Huntington's disease can take time due to communication difficulties and slower cognitive processing of information. Give adequate time for people to respond to questions, recognising that people may say things that are inconsistent. If a person's response is unclear, then explore this with them further.

Ask short, clear questions to support the processing of information without overloading the person. Avoid covering multiple issues in one assessment, as this can be overwhelming for someone with Huntington's disease. Capacity to make the decision should also be considered (more on this below).

Introducing support

When attempting to introduce new support, encourage the person to accept small amounts of help early on to make it easier for them when the condition has progressed and they require more help. Someone may initially accept help with bills, shopping or cleaning, but not personal care. If low-level support is introduced, then it can be developed into a larger care package over time.

In the early stages of Huntington's disease, people often start to need help with administrative tasks such as remembering appointments, opening letters and paying bills on time. This support, such as from a support worker or personal assistant, can help to avoid a crisis as the disease progresses. It can also establish an important relationship with a supportive person for the future.

Try to build a good therapeutic relationship before challenging the person's experience of their symptoms. For example, they may genuinely believe that they do not need help and are not experiencing symptoms of Huntington's disease, even if it seems evident that they are. They may also be frightened of their symptoms and not want to admit their extent.

When supporting a person to complete a task, break it down into smaller components and, where possible, give them an opportunity to complete the task, or components of it, themselves. Recognise that people may take longer to complete daily living activities due to slower processing. Care packages risk failing if they do not provide adequate time for a person with Huntington's disease to complete daily living activities.

Routine and consistency are important, and a care plan needs to support this. Consistency in the timing of care and the people providing care can make it easier for someone to accept essential support.

Develop detailed emergency plans that account for the behavioural and cognitive challenges of Huntington's disease, ensuring continuity of care and support during crises. Ensure carers have contingency plans in place ahead of anticipatable difficulties, as it can be difficult to develop an effective response once a crisis is happening.

Regularly review and update care plans to reflect the progressive nature of Huntington's disease and the changing needs of people and their families.

Assessing mental capacity in people with Huntington's disease

General principles

People with Huntington's disease may have impaired capacity, meaning that they are not able to make a particular decision. It should never be said that a person 'does not have capacity' in general. Capacity is decision-specific; a person might be unable to make a complex financial decision regarding where they live, but able to choose which top they want to wear today. Assume that a person has the capacity to make a decision, unless there is reason to believe they do not.

Capacity is a highly complex area - this guideline only covers specific considerations for the application of the capacity assessment process among people with Huntington's disease. The Huntington's Disease Association guideline on social work provides more detailed guidance.

A person may be deemed not to have capacity to make a particular decision for a variety of reasons. To be considered to have capacity for a decision, the Mental Capacity Act (2005) states that a person must be able to:

- Understand information that is given to them about the decision
- Retain the information long enough to be able to make the decision
- Weigh up the information available, to enable them to make the decision
- Communicate their decision in some way that is understandable to others

People with Huntington's disease may face challenges associated with each

of these components of decision-making. The role of the healthcare professional is to support them in being as involved in decision-making as possible. There are many options which can (and should) be explored to give the person the best chance of demonstrating capacity to make the decision themselves.

Due to the progression of cognitive difficulties (potentially including language difficulties, problems with speed of processing, and attentional changes), the person may not be able to *understand* information that is being given to them. Use the following strategies to maximise the likelihood of the person understanding the information being offered to them:

- Giving relevant information in small "bite-size" chunks
- Allowing sufficient time for information to be processed
- Allowing time for the person to talk
- Encouraging the person to ask questions to clarify their understanding
- Checking the person's understanding, for example, by asking them to explain what questions they have been asked
- Using favoured communication methods, such as communication boards
- Picking a time during the day where cognition is likely to be maximised. Some people with Huntington's disease sleep poorly, so they struggle at certain times of day. Talk to those who know them well, to help identify the best time for the person (where possible)

Cognitive difficulties (especially memory changes) can make it difficult for the person to retain the information they need for the decision. Remember that they only need to *retain* the information long enough to make the decision, which may be a matter of a minute or two, so memory difficulties do not by default mean that someone does not have the capacity to make a decision. To help the person retain the information, consider writing it down for them or using another visual reminder of what is being discussed.

The person may not be able to *weigh up* information sufficiently to make their decision; crucially, this requires that the person demonstrates their process of weighing up (so they need to be supported not only to actually do the weighing up, but to communicate it in some way). Think in advance of the discussion about what considerations need to be seen from the person, to be sure that they have sufficiently weighed up the decision. This would include them demonstrating that they have thought about likely

consequences of the decision in either direction, and also demonstrating their awareness of any potential serious outcomes (even if those are less likely). For example, if the person is deciding whether to begin using a wheelchair, have they considered:

- Whether their current home would enable them to use a wheelchair (are there any steps?) or whether they would have to move homes, and what that would mean for them
- How likely it is that they will fall, if they do not start using a wheelchair
- What the potential risk is to them if they do fall
- What the physiotherapist and / or occupational therapist has advised, and if the person understands why they made that recommendation

People with Huntington's disease might have specific cognitive problems, which could impair this 'weighing up' process. These include:

Lack of 'future visioning'

Some people with Huntington's disease find imagining the future difficult, which can make decision-making challenging. Present possible future consequences and test reasoning about them, rather than asking people to imagine scenarios themselves.

An ability to follow through and act on a decision:

The person may be able to say what they would do if they had a fall (e.g., 'I'd press my alarm'), but in reality would not be able to use their alarm because it is upstairs and never with them.

Communication difficulties can make it harder for a person to communicate their decision, as people with Huntington's disease may experience physical changes in their ability to speak, and cognitive changes which impact their ability to produce language. Note that 'communicating' does not necessarily just mean speech or writing; it can be as subtle as a blink, providing the parameters are sufficiently clear and agreed between the assessor and any others involved in the assessment process.

This process can require careful preparation and, where appropriate, the intervention of a speech and language therapist. Hold more than one session if needed to enable findings to be considered between sessions. This allows time for further exploration or to take into account fatigue for the person being assessed.

Involving someone who is familiar with the person with Huntington's disease, and whom they feel comfortable with, can help maximise their capacity to make a decision. Be aware that being assessed for capacity to

make a decision can feel challenging, judgemental, and even threatening. Therefore, making the process more collaborative and (ideally) pleasant and conversational is likely to help the person express themselves to their best ability.

Some people with Huntington's disease may struggle with initiating tasks as well as problem-solving. This can mean that although they may understand something in principle, being realistic about their own abilities or following through to act on a decision is difficult. Consider evidence of these factors alongside what someone may be able to communicate, as part of a mental capacity assessment. Someone may be able to talk through how they would go food shopping and the importance of eating well, and decline support with these tasks. However, if they are continuing to lose weight, do not have food at home and are requesting emergency food parcels, they may not actually be able to implement a decision in practice. Therefore, they might not be being realistic about their capabilities when they are 'weighing up' their decision.

Consider the following points from the 2018 NICE guideline on decision-making and mental capacity, where guidance is provided on assessing capacity in people with executive dysfunction:

- **Real-world observation:** Structured assessments of capacity, such as an interview, may need to be supplemented by real-world observation of the person's functioning and decision-making abilities.
- **Consulting others:** Unless it would be contrary to the person's best interests, work with paid carers, family and friends, advocates, attorneys and deputies, to explore views about the person's values, feelings, beliefs, wishes and preferences in relation to the specific decision, and to understand their decision-making history. In some cases, the views of the interested parties may differ from those of the person or the decision-maker. However, this does not necessarily mean it would be contrary to the person's best interests to consult them, and it may comprise a useful part of an assessment.
- **Making an unwise decision:** At times, the person may wish to make a decision that appears unwise. As confirmed by the third key principle of the Mental Capacity Act (2005), a person is not to be treated as unable to make a decision merely because he or she makes an unwise decision. People have the right to make a decision that others may consider unwise, provided that they have the capacity to do so. Smoking and

drinking alcohol are good examples; these choices are technically 'unwise' in terms of health and self-care. The right to make unwise decisions applies equally to people with Huntington's disease, unless they lack capacity for a specific decision.

Conclusion

Mental health symptoms and difficulties are common challenges associated with Huntington's disease. The assessment approach for people with Huntington's disease is similar to that of other people being supported by mental health services. This guideline advocates for a holistic assessment approach, considering the overlap between movement, cognition and mental health skills (see Dale et al (2022) under the References section). Not all mental health difficulties in people with Huntington's disease are 'caused' by the condition. They are often understandable responses to a highly challenging and distressing situation. Finally, Huntington's disease is a condition that affects the family, therefore assessing caregiver and family needs should form a critical part of care.

Broadly speaking, pharmacological treatments commonly used in general psychiatry can be prescribed to people with Huntington's disease effectively and safely. However, extra consideration should be given to minimise cognitive, movement and medication-related side effects. Psychological therapy might also be valued and useful – it avoids risks associated with polypharmacy, while teaching skills that people can use after therapy is concluded.

Huntington's disease care requires the input of multiple health and social care services. Co-working with local neurology services, Huntington's disease specialist services, community neurotherapy teams, social workers, the Huntington's Disease Association, and palliative care, when appropriate, is essential. Working alongside the person's family members is also important.

It is hoped that this guideline offers mental health teams the confidence and framework to support people with Huntington's disease. While the underlying condition is not yet curable, many of the mental health symptoms and difficulties associated with the disease can be treated effectively. Mental health services, therefore, have an important role to play in improving the quality of life for people with Huntington's disease.

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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/**

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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NICE guidelines are published on **www.nice.org.uk**

Relevant guidelines include:

- NICE. 2009. Depression in adults with a chronic physical health problem: recognition and management.
- NICE. 2014. Psychosis and schizophrenia in adults: prevention and management
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Notes

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