



Huntington's  
Disease  
Association

**Mental illness and  
mental capacity in  
Huntington's disease:  
A guide for mental  
health workers**

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*We'll be there*

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# Introduction

Huntington's disease is a hereditary neurodegenerative disorder. Those born naturally to a parent with Huntington's have a 50:50 chance of inheriting the gene. Most people with Huntington's develop the symptoms in their forties and fifties, although there may be subtle changes much earlier. About 5-10 % of people have onset of symptoms before age 20 (Juvenile Huntington's disease) and 10% have onset after age 60.

Huntington's disease manifests as a triad of motor, cognitive, and psychiatric symptoms which begin insidiously and progress over many years, until the death of the individual.

The average survival time after onset of symptoms is about 15 to 20 years

## The triad of symptoms of Huntington's disease

**Movement disorder** - the movement disorder is characterised both by involuntary movements (chorea), and by impairment of voluntary movements. This latter impairment often contributes more to disability than the chorea itself, resulting in reduced manual dexterity, slurred speech, swallowing difficulties, problems with balance, and falls.

Both chorea and impairment of voluntary movements progress in the middle stages of Huntington's disease, but later, chorea often declines as people become rigid and unable to initiate voluntary movements. Rigidity, spasticity and dystonia tend to emerge later in the course of Huntington's, except in cases of childhood onset, in which they are often present from the beginning. Myoclonus, is more common in juvenile-onset Huntington's disease, where it may be mistaken for a seizure. Epilepsy is uncommon, though not unheard of, in adults with Huntington's, but is said to be present in 30% of people with juvenile-onset Huntington's disease.

**Cognitive disorder** - the cognitive disorder is characterised initially by a loss of speed and flexibility. This may be seen first in complex tasks, when the person is unable to keep up with the pace and lacks the flexibility required to alternate between tasks. The cognitive disorder in Huntington's disease is considered a "subcortical" syndrome and usually lacks features such as aphasia, amnesia, or agnosia that are associated with dementia of the Alzheimer's type. The most prominent cognitive

## Introduction

impairments in Huntington's involve the so-called "executive functions" - abilities such as organisation, regulation and perception.

These fundamental abilities can affect performance in many cognitive areas, including speed, reasoning, planning, judgment, decision making, emotional engagement, perseveration, impulse control, temper control, perception, awareness, attention, language, learning, memory and timing. Cognitive losses accumulate and people develop more global impairments in the later stages of the disease. Several studies have suggested that cognitive and behavioural impairments are greater sources of impaired functioning than the movement disorder in people with Huntington's.

**Psychiatric disorder** - Psychiatric disturbances in Huntington's disease are varied. The most common specific psychiatric disorders in Huntington's are depression and anxiety.

Most people with Huntington's also experience less well defined, non-specific changes in personality and mood, such as irritability, apathy, or disinhibition as well as difficulties understanding social communication. Most of these psychiatric problems are believed to be related directly to the central nervous system injury caused by Huntington's. People with Huntington's who have psychiatric disorders generally suffer from under diagnosis and under treatment. It is important to remember that psychiatric problems, particularly depression, are very common and devastating in Huntington's, but they are also treatable. Relieving a depression in someone with Huntington's may be the single most effective intervention a physician can perform.

## Research

Although we can't currently cure Huntington's disease, we can treat many of the symptoms. There is also a lot of hope for the future in terms of finding better treatments and hopefully something that will delay the onset of the condition. For more information on research in Huntington's see the HDBuzz website [en.hdbuzz.net](https://en.hdbuzz.net)

## Types of mental illness in Huntington's disease

Mental illness is very common in Huntington's disease. Sometimes people with Huntington's have found it hard to access support for their mental illness from mental health services as the problem is said to be "physical and not mental". However, mental illnesses are often caused by physical brain problems and this should not lead to exclusion from mental health care.

### **Organic personality disorder (ICD 10 code F07.0)**

This is a mental illness which is characterised by the following symptoms;

- **Inability to persevere with goal-directed activities**
- **Altered emotional behaviour including irritability, emotional shallowness and apathy**
- **Expression of needs and impulses without the consideration of others**
- **Excessive pre-occupation with single themes**
- **Altered sexual behaviour**

If two or more of the above are present and the problem is caused by a brain disorder (including head injury or Huntington's disease) then this diagnosis may be made. This constellation of symptoms is very common in Huntington's. This diagnosis is no less important than schizophrenia and bipolar disorder. Legal bodies consider organic personality disorder in exactly the same way as other mental illnesses (for instance in the use of the Mental Health Act).

### **Dementia in Huntington's disease (ICD 10 code F02.2)**

This diagnosis is usually reserved for people with Huntington's who have problems with cognition over a number of areas, including memory. This refers to a "global impairment of cognitive function" and is usually only present in later stages of Huntington's.

### **Delirium (ICD10 code F0.5)**

This diagnosis usually refers to a sudden reduction in orientation and awareness and should be a sign for further investigation for additional problems such as infections or subdural haematoma. A sudden change in orientation and awareness is not at all typical or the progression of Huntington's on its own.

### **Other diagnoses**

People with Huntington's are at increased risk from a variety of mental illnesses and are also at risk for suicide and self-neglect. Common mental illnesses include generalised anxiety disorder, panic disorders, phobias, depression and substance misuse. Psychosis is relatively rare in Huntington's but does occur. Sometimes it is difficult to tell what part the Huntington's played in the causation of these illnesses but they can often be treated. People with Huntington's disease have a right to be seen and treated by mental health services.

## Treating mental illness in Huntington's disease

There is nothing about Huntington's disease that prevents the treatment of any mental illness. As a general principle, people with depression, anxiety and psychosis can be treated in the same way as people with those illnesses but no Huntington's. However, there are a number of treatments for organic personality disorder which are effective but depend on the symptoms being treated. There are a number of specialist clinics for people with Huntington's in England and Wales, most have a Neuropsychiatrist linked to them which can be invaluable in the treatment of Huntington's.

### **Drugs for chorea and mental illness**

A number of drugs which are used to treat chorea can cause depression, anxiety and cognitive impairment. Drugs such as tetrabenazine, risperidone, sulpiride and olanzapine can all lead to these symptoms sometimes. A good rule is to consider whether or not the anti-chorea drugs are actually needed and to stop them if they are not.

### **Treating organic personality disorder**

Educating family members and caregivers about the personality changes seen in Huntington's can help them to change the environment, which, in turn, might lessen symptoms. Reducing demands on the person with Huntington's and keeping to regular routines can often improve irritability. Irritability has a number of causes and these need to be clearly investigated (for instance pain, infection, hunger or cognitive overload). These causes will lead to different treatments. When other underlying causes of irritability have been ruled out and psychosocial interventions have been utilised, there are a number of medications that have been used to treat irritability.

Selective serotonin reuptake inhibitors (SSRIs) are commonly used in this situation but other drugs such as lamotrigine, valproate, carbamazepine and mirtazapine have also been used. Nabilone, a synthetic cannabinoid, helped irritability and anxiety in a small pilot study. Apathy is common in Huntington's but is rarely complained of by people with Huntington's and is difficult to treat. Often, managing

## Treating mental illness in Huntington's disease

the expectations of carers and family members can be the most effective intervention. Excessive pre-occupation with single themes, or perseveration, can sometimes also respond to SSRIs.

### **Treating depression and anxiety**

Standard pharmacological approaches to the treatment of depression and anxiety should be used. However, be aware that cognitive overload may be a significant maintaining factor and, if this can be lessened, it will improve the chances of remission. Changes in physical function, for instance choking, may lead to eating phobia and then to depression; therefore managing the swallow may again be a key to remission. People with Huntington's and depression/anxiety commonly respond well to treatment.

### **Psychosis**

Psychosis is rare in Huntington's and, where it occurs, it is more often resistant to treatment. The standard process for managing psychosis should be employed.

### **Delirium**

Be aware of the possibility of delirium in people with Huntington's. They might have a low threshold for testing for underlying causes, particularly chest and urinary infections and subdural haematoma. This is readily treated if diagnosed early.



## The Mental Capacity Act (MCA) 2005 and its guiding principles

A person with Huntington's disease cannot be globally assessed as 'not having capacity'. The Mental Capacity Act makes it clear that: The assessment of capacity must be about the particular decision that has to be made at a particular time and is not about a range of decisions;

- **If the person with Huntington's cannot make complex decisions, this does not mean that he/she cannot make simple decisions;**
- **People with Huntington's may lack capacity in specific areas. The mental incapacity may be permanent or temporary.**

### The key principles in relation to Huntington's

#### Key principle 1

A person must be assumed to have capacity unless it is established that they lack capacity.

It cannot be decided that someone lacks capacity because they have Huntington's or because of the way they behave.

#### Key principle 2

A person is not to be treated as unable to make a decision unless all practical steps to help them to do so have been taken without success.

The specific cognitive challenges that people with Huntington's must be taken into account (See the sections on assessing capacity and cognitive impairment in Huntington's).

#### Key principle 3

A person is not to be treated as unable to make a decision merely because he makes an unwise decision.

Some of the decisions that an individual takes may be seen by others as unwise. For example the decision to buy a £30,000 car which will use all of their savings meaning that they won't be able to pay their rent and have to move out. Most people would see this as unwise, but this does not mean that the person does not have capacity. If they are able to give clear reasoning for their actions (they may say they had decided they actually need a smaller place anyway; they know that they won't be able to drive soon because of Huntington's but they love cars and this is their last chance to ever do this, they have thought about their driving skills and have informed the DVLA that they have early symptoms and are being tested each year etc.).

## Assessing capacity in Huntington's disease

The starting point for any test of capacity is an assumption that the person with Huntington's disease has capacity. If, however there is doubt, then they must undergo the two-stage test of capacity as follows:

- **Is there an impairment of, or disturbance in the functioning of a person's mind or brain?**
- **Is the impairment or disturbance sufficient that the person lacks the capacity to make a particular decision?**

If the first stage of the test of capacity is met, the second test requires the person to show that the impairment or disturbance of brain or mind prevents them from being able to make the decision in question at that time and is assessed using The Functional Test.

### The Functional Test

The test focuses on how the decision is made, rather than the outcome or the consequence of the decision.

- **To understand the information relevant to the decision**
- **To weigh that information as a part of the process of making a decision**
- **To communicate his/her decision**

This test must be complete and recorded; the documentation must demonstrate the above process.

### General principles when assessing a person with Huntington's using the two-stage test of capacity

People with Huntington's may have impaired capacity for a number of reasons. The primary reason is related to the inability to "weigh up" information sufficiently to make a decision. Communication difficulties can make it difficult for a person to express the decision making process but communication problems should rarely be the prime reason for incapacity. The following strategies can be used to maximise communication:

- **Giving information in "bite-size" chunks**
- **Allowing time for information to be processed**
- **Allowing time for the person to talk**

- **Using favoured communication methods (for instance communication boards)**
- **Picking a time during the day where cognition is greatest**

This process might require careful preparation and, where appropriate, the intervention of a speech and language therapist. More than one session may be required to enable material to be digested between sessions. Involving a person who is familiar with the person with Huntington's could help maximise the chance of reaching a capacitous decision.

### **Weighing up the pros and cons**

Deciding how much "weighing up" is sufficient for capacity is tricky and the law is not clear on this matter. It is important to make an estimate before the assessment about how much "weighing up" would be sufficient in which case. This usually means that the person being assessed should consider both the common and the serious consequences of a decision. People with Huntington's might have specific cognitive problems which could impair this process.

1. Perseverative thinking - many people with Huntington's will find it difficult to reflect on the negative consequences of a decision that they have made; they just "want what they want". However, if presented with potential negative consequences, they can often show good reasoning around practical solutions to the problems. Most people agree that this constitutes a capacitous decision.
2. Lack of "future visioning" - some people with Huntington's find imagining the future difficult. This might make capacitous decision-making difficult in relation to imagining the consequences of decisions. As is the case with perseverative thinking, it might be appropriate to present possible future consequences and to test reasoning about them rather than asking people to spontaneously generate the scenarios themselves.

### **Do people with Huntington's disease have a "mental disorder" as defined by the Mental Capacity Act?**

The vast majority of people with Huntington's disease have a mental disorder as defined by the Act. It would be exceedingly rare to be asked to do a capacity assessment on a person with Huntington's disease who does not have a mental disorder (see section on mental illness).

**Important note 1:** A cognitive test (such as the mini-mental test) is not a test of capacity; the two stage test must be used.

**Important note 2:** Decision makers - this does not have to be a Doctor. It is whoever the person is making the decision that another individual does not have the capacity to make a particular decision. This could be a relative or paid carer (when deciding what meal to have that day), a Dentist (when a tooth is to be removed), an Occupational Therapist (when deciding what equipment is required for safe transfers) etc. or a social worker in the case of a decision about social care.

**Important note 3:** There can be circumstances where the person with Huntington's gives a false impression to the assessor. They may present incredibly well when their capacity is being assessed and give false answers. For example, an individual who is being assessed in relation to their ability to make a decision about living at home may tell the assessor that they are able to shop independently and prepare nutritious meals for themselves three times a day. The person who has raised concerns may say that actually the furthest they get is the local café where they bring home cream cakes and this is what they live off. The assessor needs to take into account information which has been provided to them by others and ask questions of the individual that will highlight any lack of insight (e.g. can you describe how you get to the shop/how you prepare the meal).

## Best interest

The law requires that where decisions are being taken for/about a person who is not able to make that decision themselves, then it must be made in their best interest.

The Mental Capacity Act sets out the principle of best interests as follows:

### Key principle 4

An act done, or decision made, under this Act for or on behalf of a person who lacks capacity must be done, or made, in their best interests.

### Key principle 5

Before the act is done, or the decision is made, regard must be had to whether the purpose for which it is needed can be as effectively achieved in a way that is less restrictive of the person's rights and freedom of action.

A person trying to work out the best interests of someone who lacks capacity to make a particular decision ('lacks capacity') should:

#### Encourage participation

- Do whatever is possible to permit and encourage the person to take part, or to improve their ability to take part, in making the decision.

#### Identify all relevant circumstances

- Try to identify all the things that the person who lacks capacity would take into account if they were making the decision or acting for themselves.

#### Find out the person's views

- Try to find out the views of the person who lacks capacity, including:
  - the person's past and present wishes and feelings (these may have been expressed verbally, in writing or through behaviour or habits.)
  - any beliefs and values (e.g. religious, cultural, moral or political) that would be likely to influence the decision in question
  - any other factors the person themselves would be likely to consider if they were making the decision or acting for themselves

#### Avoid discrimination

- Don't make assumptions about someone's best interests simply on the basis of the person's age, appearance, condition or behaviour.

### **Assess whether the person might regain capacity**

- Consider whether the person is likely to regain capacity (e.g. after receiving medical treatment). If so, can the decision wait until then?

### **If the decision concerns life-sustaining treatment**

- Don't be motivated in any way by a desire to bring about the person's death. They should not make assumptions about the person's quality of life.

### **Consult others**

- If it is practical and appropriate to do so, consult other people for their views about the person's best interests and to see if they have any information about the person's wishes and feelings, beliefs and values. In particular, try to consult:
  - anyone previously named by the person as someone to be consulted on either the decision in question or on similar issues
  - anyone engaged in caring for the person
  - close relatives, friends or others who take an interest in the person's welfare
  - any attorney appointed under a Lasting Power of Attorney or Enduring Power of Attorney made by the person
  - any deputy appointed by the Court of Protection to make decisions for the person.
- For decisions about major medical treatment or where the person should live and where there is no-one who fits into any of the above categories, an Independent Mental Capacity Advocate (IMCA) must be consulted.
- When consulting, remember that the person who lacks the capacity to make the decision or act for themselves still has a right to keep their affairs private – so it would not be right to share every piece of information with everyone.

### **Avoid restricting the person's rights**

- See if there are other options that may be less restrictive of the person's rights.

### **Take all of this into account**

- Weigh up all of these factors in order to work out what is in the person's best interests.

## Restraint

Restraint is defined in Section 6 of the Mental Capacity Act as:

- **the use or threat of force to secure the doing of an act that the individual resists; or**
- **the restriction of the individual's liberty whether that individual resists or not**

Restraint or restrictions on an incapacitated individual's liberty can be justified under the Mental Capacity Act provided:

- **reasonable steps are taken to establish that the individual lacks capacity in relation to the matter in question; and**
- **it is reasonably believed that the individual does lack capacity in relation to the matter in question; and**
- **it is in the best interests of that individual for the act to be done; and**
- **it is reasonably believed that it is necessary to do the act to prevent harm to that individual; and**
- **the act in question is a proportionate response to the likelihood of the individual suffering harm; and**
- **the act in question is a proportionate response to the seriousness of that harm**

However, the distinctions between restraining or restricting an individual on the one hand and depriving them of their liberty on the other, are not always easy to identify.

For example, it is possible to 'deprive someone of their liberty' not just by physical confinement, but also by virtue of the level of control exercised over a person's movements. A high level of control over someone, such as controls over who can visit them, and when they can conduct certain activities, may result in a finding that they are being deprived of their liberty.

The concepts of restraint, restriction and deprivation of liberty are best understood as existing on the same 'spectrum of control', with deprivation of liberty involving a higher degree or intensity of control over that individual. Ultimately, the concept is one to be interpreted in view of the specific circumstances of that individual.

**Whatever the situation, a Deprivation of Liberty is unlawful unless authorised.**

## Deprivation of Liberty Safeguards (DoLS\*)

The DoLS procedure aims to 'safeguard' the liberty of a person with Huntington's who has been deemed not to have capacity in relation to where they reside by ensuring that a rigorous and transparent procedure is followed prior to any deprivation of liberty. The DoLS procedure is aimed at ensuring that those caring for, or involved with, incapacitated individuals are able to engage with decision-making involving questions about their liberty. The DoLS procedure is also aimed at ensuring that such decision-making is conducted carefully, and is subject to independent scrutiny.

While there may be extreme cases where an individual with Huntington's has to go through the DoLS procedure in many cases, once the person is in an appropriate placement, they are happy to reside there. Whilst the thought of a residential placement can seem daunting and upsetting (especially for someone with Huntington's who struggles with 'future visioning' and may have negative memories from other family members) if the person is well fed, appropriately medicated, empowered to participate in activities that they enjoy and cared for by people who understand them and their condition then they usually begin to enjoy the security of a placement. Most residential placements who care for people with Huntington's would offer an open door policy where there is space for people to roam into a garden or secure area; a locked door can cause problems.

For information on homes that care for people with Huntington's contact us at the Huntington's Disease Association.

In addition, there is scope for the person to be legally represented. Anyone who does not have family or friends who can be consulted will be referred to an Independent Mental Capacity Advocate (IMCA) who will be instructed to support and represent them during the assessment process. An IMCA is a specialist advocate trained to advise and represent people in relation to mental capacity issues.

\*DoLS replacement bill approved by Parliament with Liberty Protection Safeguards (LPS) due to come into force in 2020. This is likely to run alongside DoLS for 12 months for transition to LPS.

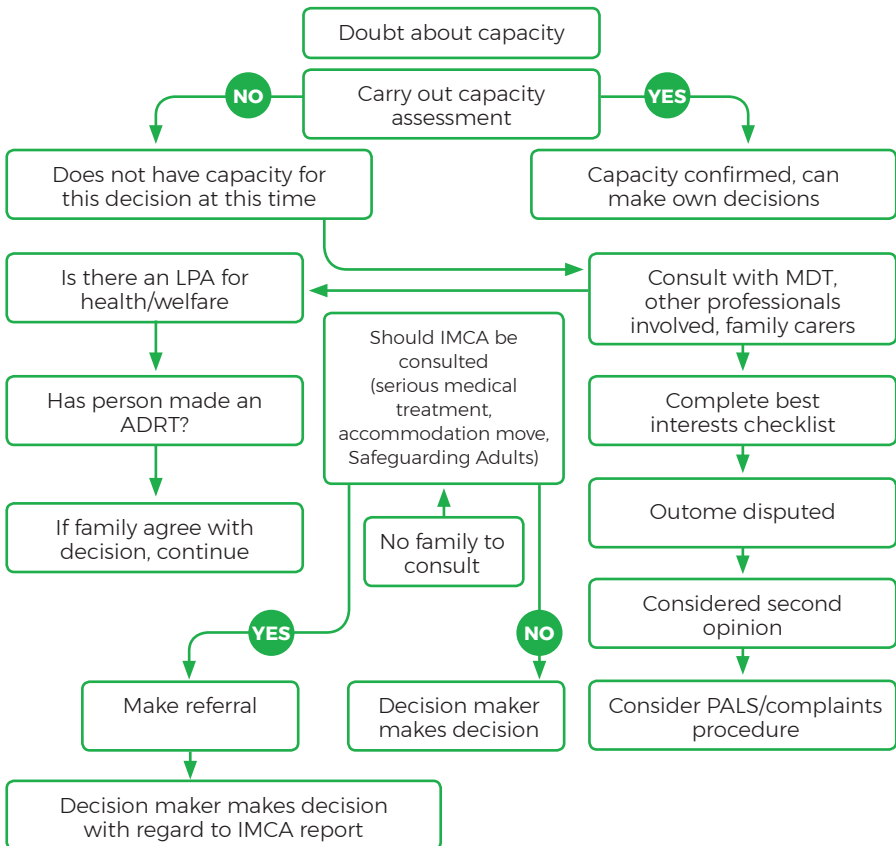


## Mental Capacity Act and Mental Health Act Pathways

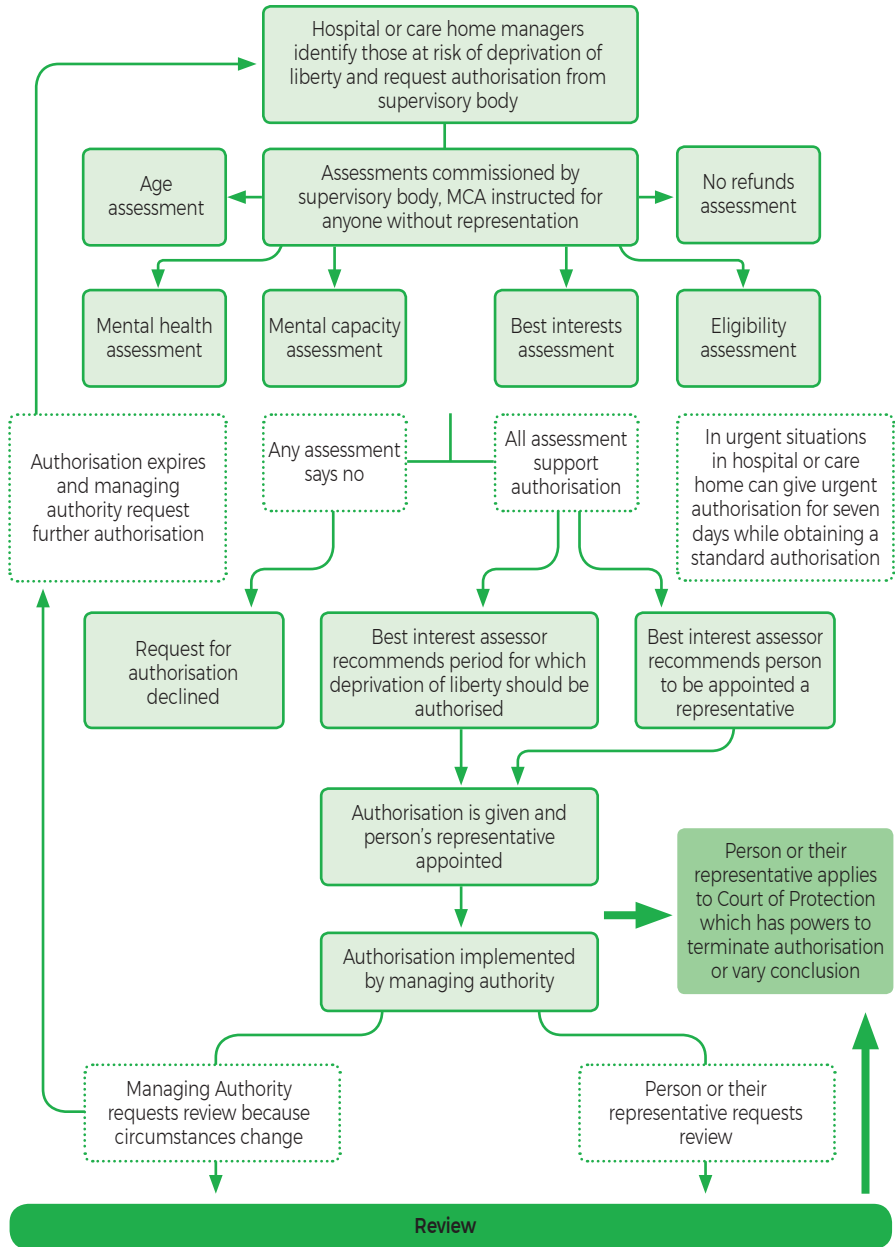
All adults affected by Huntington's, should be presumed to have capacity unless the opposite has been demonstrated. Consent must be obtained by the person undertaking the procedure and is specific to the decision to be made.

The diagram set out below should assist as to what pathway would need to be followed, ensuring that the guiding principles referred to in Section 3 are followed at all times.

### Mental Capacity Act Pathway



## Overview of the Deprivation of Liberty Safeguards process

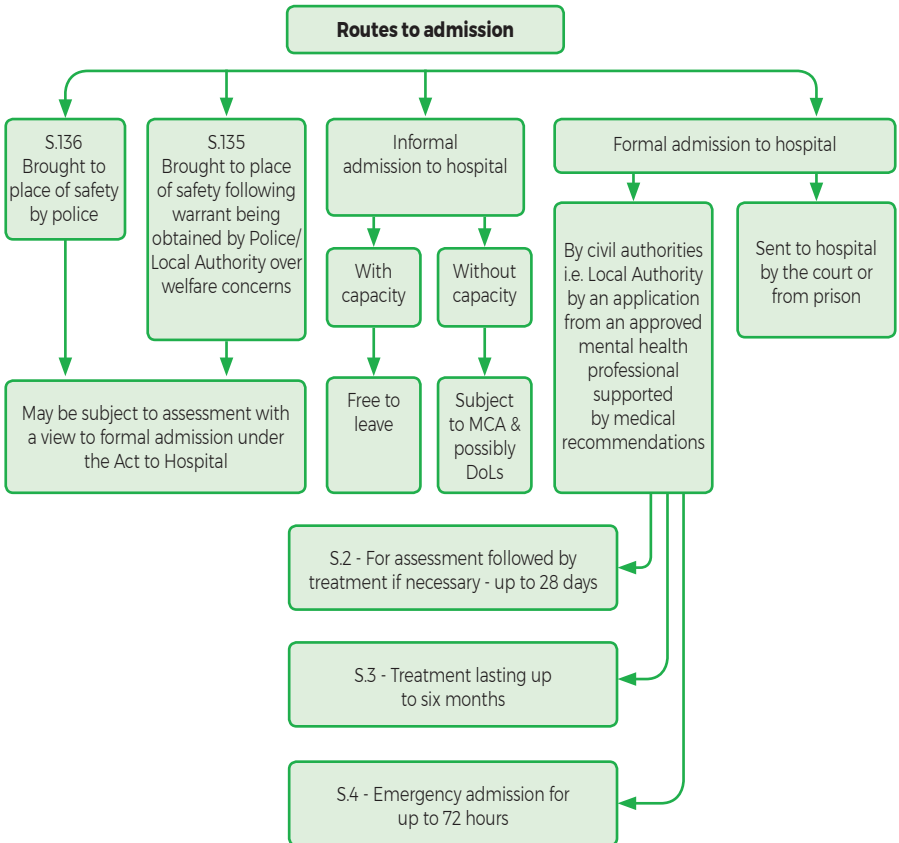


## Mental Health Act Pathway

Consideration may also need to be given with regard to whether or not the Mental Health Act applies. This may consider if the person may need to be assessed as to whether they should receive some assistance at home for their mental health condition or in hospital, which may be against their wishes.

There are different routes and options available, the most common of which is set out in a pathway diagram below. What is important to remember is that some of the symptoms of Huntington's are well recognised forms of mental illness that subsequently present a form of risk to the person, their health or safety, or to others.

## The main Mental Health Pathways



## Discharge planning

If a person has been in hospital, as a result of physical or mental illness, good discharge planning is essential. If the person is going home, the following should be considered:

- **Can the family cope with having them at home again?**
- **Think about what preceded the admission and if this is now resolved / measures put in place to prevent this happening again**
- **What local support has been put in place (e.g. ongoing involvement of CMHT)?**
- **Does the person and their carer know who to contact if things are going wrong?**
- **Does the person and / or their carer know how any new medications will be accessed?**
- **If a new care agency is being put in place, have they had adequate training?**

If a residential setting is now needed it is important to know that they will be able to cope with the person. At the Huntington's Disease Association, we have a list of homes that have experience of Huntington's and are happy to work with homes to provide training. Make sure that all relevant information has been passed to the home including essential information about behavioural triggers.

## Adult safeguarding

It is important to emphasise that those who need safeguarding help are often people who due to age, disability, incapacity, physical or learning disabilities and those with mental health needs who are at risk of suffering harm both in institutions and in the community.

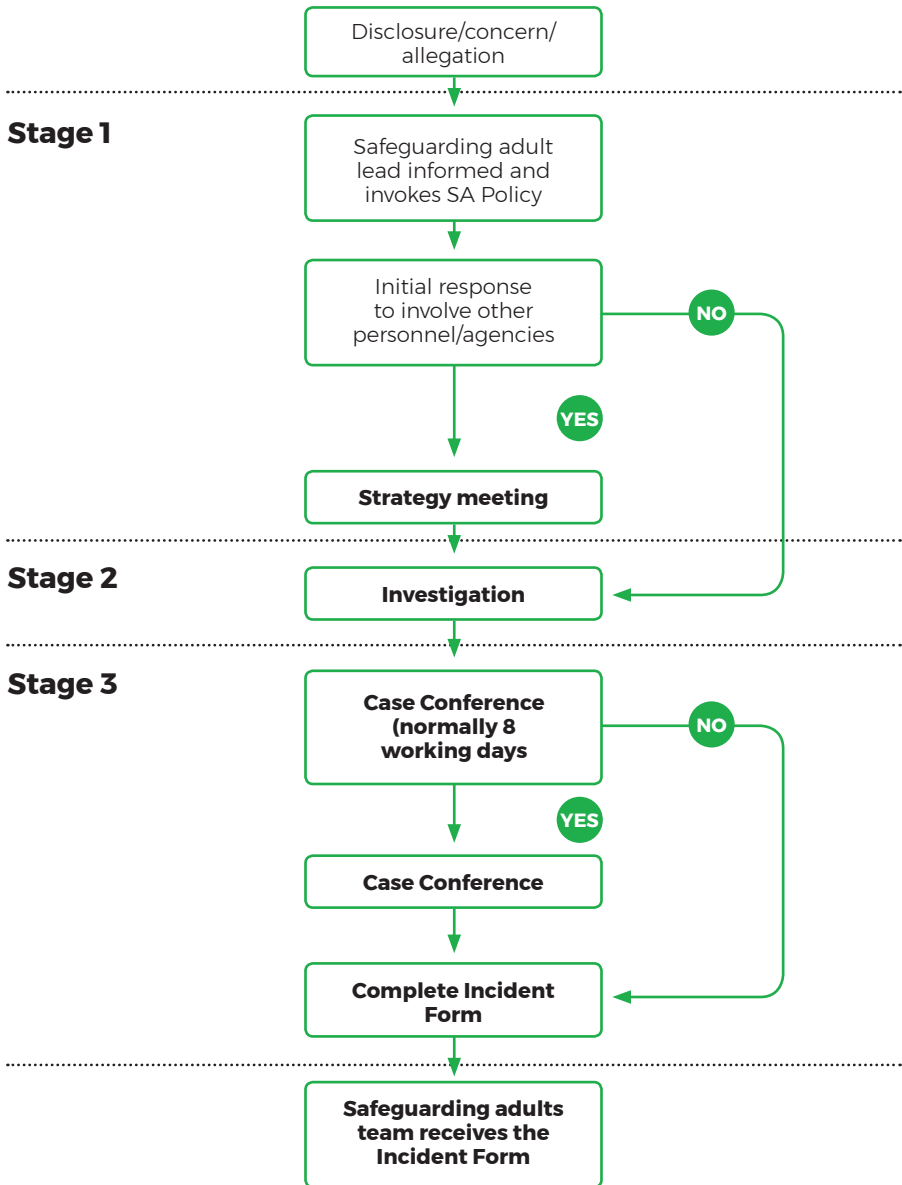
The six key concepts are embedded in The Care Act 2014:

1. **Empowerment**
2. **Protection**
3. **Prevention**
4. **Proportionate responses**
5. **Partnership**
6. **Accountability**

Health and Social Care organisations have an important role in the protection of members of the public from harm and are responsible for ensuring that services and support are delivered in ways that are high quality and safe. At the same time, they are also responsible for conducting safeguarding enquiries that are highlighted, but must do so lawfully, taking account of best interests, along with the rights of all those involved, in particular, individuals, families and carers.

A generic overview of the safeguarding process is set out overleaf.

## Generic flowchart - Implementing Safeguarding Adults policy



## The rights of individuals, families and carers

We have summarised a number of pathways that may be followed for a person with Huntington's because of them presenting in a particular way which has highlighted concerns for the family or for professionals involved in providing health or social care support. This means that decisions will need to be taken about what may happen next for the person, common examples being:

- **What support they may require to live at home**
- **Whether they may need assistance to take medication**
- **Whether they are able to continue to manage their own money and affairs**
- **Whether they can keep themselves safe at home or from others because of possible abuse over money, possessions, personal security etc.**

Often when these types of situations arise, there is agreement between all concerned, e.g. family members, close friends, the local authority (most often in the form of a social worker or care coordinator), about what should happen, and there is involvement of those parties to arrive at a decision that is in the best interest of the person affected.

A number of high profile cases that have been before the Courts since 2009, along with investigations by the Care Quality Commission and the Equality and Human Rights Commission, have highlighted that serious mistakes have been made by health and social care professionals when dealing with individuals, who for various reasons have lost the capacity to make decisions for themselves.

In some cases a person was deemed to lack capacity because their decision was seen unwise or it was thought that they needed protection because of perceived vulnerability. Given this, it is crucial that there is clarity about the rights of:

- **Individuals**
- **Families**
- **Carers**

Equally, it is crucial that health and social care professionals are clear as to their local Best Interest and adult safeguarding processes.

## The rights of the individual

What is crystal clear from the legislation and as highlighted above, is that individuals do have rights, and that steps should be taken to support people who have difficulties that may arise from a progressive condition such as Huntington's. The key rights are enshrined in the statutory principles referred to in section 3.

## The rights of the family

Family members do have rights, most critically to be consulted about decisions that may need to be taken.

One of the big difficulties for families is where they have cared for a person for many years without any involvement from professional bodies, making most if not all the decisions for their relative. However, the family along with agencies that have since been engaged must observe and respect the rights of the individual, and where decisions need to be taken, they need to be implemented on the basis of best interest. Within this process, families have a right to be involved and consulted and appropriate weight given to their views.

What is important for family members to consider in respect of a relative who has a diagnosis or who shows symptoms of Huntington's, is to take the opportunity to forward plan the way the family member can lawfully secure more decision making powers when the condition of the relative with Huntington's deteriorates to the point that they are no longer able to make decision for themselves.

One important distinction to the involvement of family members, is where the local authority is proposing to make an application for the relative to be detained under the Mental Health Act for Treatment. In this instance, the worker proposing to make the application to a hospital is legally bound to establish whether the nearest relative objects to the application being made. Where this is the case, and the nearest relative (Note: this is defined by law and sometimes changes due to relationships, caring roles etc) does object, the person cannot be admitted until the nearest relative has been displaced by the local authority obtaining an order displacing them as such.



## The rights of carers

These are very similar to those set out for family members, not least where the carer may have had a notable involvement with the individual affected, and because of this, their views must be taken into account, even where they may have been accused of not having looked after a person appropriately

## Cognitive impairment in Huntington's

Cognitive impairment in Huntington's is probably one of the most disabling features of the illness, but because earlier on in the illness some of the changes are very subtle, it can often be overlooked. Cognition refers to the mental processes including attention, remembering, producing and understanding language, solving problems and making decisions.

It is significant that the frontal lobe is affected in Huntington's. This has responsibility for organising, prioritising, controlling impulses, monitoring self awareness, motivation, finishing tasks, creative thinking and problem solving. This can lead to a variety of challenges when treating someone with Huntington's. They may not have an awareness that they have a difficulty in a particular area, and perceive that they are managing well, when in fact this is not the case.

Careful assessment of situations is needed and if possible seeking the view of a carer may give a clearer picture of the actual situation.

### **Five key changes to cognition are as follows:**

**Slowed thinking** is one of the most disabling features of the cognitive impairment. It occurs because there are fewer brain cells working which slows down the processing of information which results in slower thinking and responses being slower. This is extremely tiring and frustrating for the person with Huntington's and may lead to irritability.

**Difficulty in switching** or maintaining focus results in a difficulty in multi-tasking. This may be even simple things like watching television and being asked a question, or walking and talking. It may result in the person persistently dwelling on the same thing, or becoming irritable/ having an outburst of temper for no apparent reason. Understanding this and adapting the environment can be helpful.

**Recognition** is easier than recall, this means that a person's memory is not necessarily affected by Huntington's. The problem that people have is accessing those memories. This can be helped by asking questions that narrow down the answer and give options to choose from, like prompts.

A person may get stuck or focus on a particular topic, this is called **perseveration**. There are a number of ways that the person can be helped with this including distraction, setting boundaries by explaining this has already been discussed or setting a time amount for the length of time a subject will be discussed with them.

Cognitive impairment affects a person's ability to motivate or complete a task It also affects their ability to organise and plan things. This can result in chaotic organisation or the inability to change a course of direction of thinking. We often make snap decisions, subconsciously thinking through the options before deciding on something, and this becomes impossible in Huntington's. It means thoughts that used to be automatic now have to be conscious which makes it difficult for the person.

Huntington's affects a person's ability to understand the wider picture around them. This in turn affects their ability to wait for anything. They can't see that there are other pressures that may make it difficult to respond to things immediately.

Florence Nightingale cited

*“apprehension, uncertainty, waiting, expectation, fear of surprise do a patient more harm than exertion.”*

This is a good summary of the effects of the cognitive difficulties in Huntington's.

## Forward planning in advanced Huntington's disease

There can be little doubt that significant life events – from redundancy, ill health, incapacity, and through to death itself are much better dealt with if there has been some forward planning. Here are a few measures that can be considered and taken.

### Wills

At its most basic, a Will lets the individual decide what happens to their money, property and possessions after their death.

A person can write a Will themselves, but they should get legal advice to make sure their Will is interpreted in the way they wanted. The person needs to get their Will formally witnessed and signed to make it legally valid. If they want to update their Will, they need to make an official alteration (called a codicil) or make a new Will. If a person makes a Will they can also make sure they do not pay more Inheritance Tax than they need to.

### Lasting Powers of Attorney (LPA)

Enduring Powers of Attorney have been replaced by Lasting Powers of Attorney in England and Wales. However, they can still be used if they were made and signed before October 2007.

A Lasting Power of Attorney is a legal document that lets the individual (the 'donor') appoint people (known as 'attorneys') to make decisions on their behalf. LPAs are designed to be recognised by financial institutions, care homes and local authorities, as well as tax, benefits and pension authorities.

They are legal documents that can be set up relatively cheaply. The donor must be 18 or over and have mental capacity - when making their lasting power of attorney. It could be used if a person became unable to make their own decisions.

There are two types of Lasting Power of Attorney (People can choose to make one type or both)

- **Health and welfare property**
- **Financial affairs**

The Office of the Public Guardian ([publicguardian.gov.uk](https://www.publicguardian.gov.uk)) help to

protect people who lack capacity. Their role includes setting up and managing a register of Lasting Powers of Attorney.

### **Advance Decision to Refuse Treatment (ADRT)**

An ADRT is a decision a person can make now to refuse a specific type of treatment at some time in the future. It allows their family, carers and health professionals know whether they want to refuse specific treatments in the future. This is so a person's wishes are known if they are unable to make or communicate those decisions at the time it is required.

The treatments a person is deciding to refuse must all be named in the advance decision. Sometimes, they may want to refuse a treatment in some situations but not others. If this is the case, they need to be clear about all the circumstances. They can refuse a treatment that could potentially keep them alive ('life sustaining' treatment). This includes treatments such as ventilation and Cardio Pulmonary Resuscitation.

They may want to discuss this with a doctor or nurse who knows about their medical history before they make up their mind.

A person can make the ADRT, as long as they have the mental capacity to make such decisions. They may want to make an advance decision with the support of a clinician.

At the Huntington's Disease Association, we have created forms for making advance decisions which are available on request.

Is an advance decision legally binding? Yes it is, as long as it: complies with the Mental Capacity Act; is valid; applies to the situation.

#### **An advance decision may only be considered valid if:**

- **The individual is aged 18 or over and had the capacity to make, understand and communicate their decision when they made it**
- **They specify clearly which treatments they wish to refuse**
- **They explain the circumstances in which they wish to refuse them**
- **It is signed by the individual and by a witness if they want to refuse life-sustaining treatment**
- **They have made the advance decision of their own accord, without any harassment by anyone else**

- **They have not said or done anything that would contradict the advance decision since they made it (for example, saying that they have changed their mind)**

As long as it is valid and applies to the person's situation, an advance decision gives the health and social care team clinical and legal instructions about the person's treatment choices. An advance decision will only be used if, at some time in the future, the person is not able to make their own decisions about their treatment.

### **Deputy appointment**

A deputy is someone appointed by the Court of Protection to make decisions for someone who is unable to do so on their own. The Mental Capacity Act is used to work out if someone can make their own decisions and how they can be helped. The court will not appoint someone as a deputy if the person is able to make their own decisions. A relative may be able to make a lasting power of attorney instead (see above).

A deputy is responsible for making decisions for someone until either the person they are looking after dies or is able to make decisions on their own again.

### **Appointee**

A person can apply to the Department for Work and Pensions (DWP) for the right to deal with the benefits of someone who cannot manage their own affairs because they are mentally incapable or severely disabled. Only one appointee can act on behalf of someone who is entitled to benefits (the claimant) from the DWP. An appointee can be: an individual, e.g. a friend or relative, an organisation or representative of an organisation, e.g. a solicitor or local council.

The DWP arranges to visit the claimant to assess if an appointee is needed. The DWP interviews the individual to make sure they are a suitable appointee. During the interview, the individual and the interviewer fill out an appointee application form (Form BF56). If DWP agrees with the application, Form BF57 (confirming the individual has been formally appointed to act for the claimant) is issued. The person is not the appointee until this happens. Once authorised, DWP will monitor the situation to make sure it is still suitable for the appointee and the claimant.

## JHD and Parental Responsibility

Juvenile Huntington's disease (JHD) is a less common form of the disease that starts to manifest in young people under the age of 20 years.

The law imposes various age limits in respect of some legislation, and not for others. Why is this important? Because it determines who may be able to make decisions about an individual. This becomes of significant importance when dealing with people under the age of 16. Why? Because the law lays down the following age limits for the following legislation:

- **Mental Capacity Act - 16 years and over and Deprivation of Liberty Safeguards - 18 years and over**
- **Mental Health Act - 0 years - until death (So no age limit)**

### **So what about people under 16?**

The general rule is that for those under 16, decisions for or about them are made by one or more persons who have what the law terms 'Parental Responsibility'.

### **Who has Parental Responsibility? (PR)**

A mother automatically has parental responsibility for her child from birth. A father usually has parental responsibility if they:

- **are married to the child's mother at the time of birth**
- **are listed on the birth certificate (after a certain date, depending on which part of the UK the child was born in)**
- **subsequently marry the mother**
- **apply to the Court for an order in respect of Parental Responsibility**

In addition, there are other situations when Parental Responsibility can be obtained, for example:

- **Where a person is adopted**
- **Where a person under 16 is taken in to the care of the local authority by means of an interim or full care order. In such situation, Parental Responsibility is shared with one or more parents who have Parental Responsibility**

- **Where a Residence Order has been obtained from the family courts, for example by a relative following the separation of the parents and there have been difficulties over that relative being able to have contact with the child relative, e.g. grandparents.**

But what if the young person does not agree with the parent's (or persons with Parental Responsibility) decision about them?

In this case, 'competence' of the young person would be considered. This means that the age, understanding and ability of the young person to understand the issues and the complexities would be taken into account. This has happened and the Courts have supported young people to be able to make their own decisions.



## Day to day management of a person with Huntington's

### Communication

Due to the cognitive and motor difficulties it is necessary to give people with Huntington's time and patience when communicating. People with Huntington's need time to receive the message, process the information and answer.

Often the care giver has to take responsibility for initiating and to maintain conversation.

Communication problems in Huntington's occur because the muscles cannot move with the correct range of movement, speed, force or coordination. The first signs of speech problems usually begin to appear as slurred or slowed speech.

Involuntary movements may affect the muscles involved in breathing, word forming, respiration and articulation. There may be changes in the way someone with Huntington's speaks e.g. prolonged spaces between words or syllables and the person may lack variation in pitch.

Reduced initiation of communication, reduced vocabulary, word finding problems and difficulty in understanding more complex or subtle aspects of language mean that it is necessary to keep communication simple.

The emotional changes that often manifest in Huntington's e.g. depression, personality changes such as irritability, lack of motivation can have an impact on communication. As Huntington's progresses to the mid stages these continue and people find it difficult to stay on topic. They may repeat themes, topics, thoughts or ideas (perseveration).

Communication is usually fully compromised and may be non-verbal by the later stages but the person can usually understand simple instructions and conversation. It is good practice to introduce low tech equipment at an early stage before reading, writing and speech is impaired.

## Tips to aid good communication

- Speak slowly and clearly but not in a patronising way
- Allow time and repeat or rephrase if necessary
- Provide cues to help find the right words
- Don't hurry a conversation
- Simplify the message with closed questions
- Reduce the number of choices
- Communicate for short periods
- Reduce distractions and noise levels.
- Monitor listening ability

## Further tips

**Compliance:** It is very difficult for people with Huntington's to make changes to their routine and do 'new things'. Often they will refuse to engage with services, especially if they have been newly referred. This should not be seen as a reason to stop services; if anything it shows that they are even more in need. Home visits may be an option (people are more likely to respond to this than having to travel to an appointment). Even on a home visit professionals may find that their entry may be refused. It is advised to talk to family and establish when would be a good time to go, how they might best engage or plan a joint visit with a professional who is already engaged. If this person is in denial they aren't going to understand the situation. Sometimes there may be no way of engaging initially so professionals may need to work with family and friends to support them in their caring role and work together to best support the person.

**Food and drink:** Weight loss is an ongoing challenge to people with Huntington's. People with Huntington's may need more calories per day to maintain their body weight. People with Huntington's tend to have ravenous appetites and can be hungry all of the time. If people are underweight, they need to eat little and often throughout the day (and night). It is advisable to weigh the individual at regular intervals. Advice should be taken from a dietitian.

**Choking:** As Huntington's worsens, people develop a swallowing disorder. It may not be obvious that the person is having difficulty swallowing, so it is important to monitor and watch whilst they are eating to prevent choking. People with Huntington's have a tendency

to eat too fast, forget to chew or cram food into their mouths. This increases the risk of choking. Advice should be taken from a Speech and Language Therapist.

**Falls:** Huntington's affects people's mobility and balance. Close attention should be paid to walking and transfers to prevent falls. Advice should be taken from a physiotherapist.

**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 look unhappy, bored or uninterested. Weakness and changes in posture (such as leaning to one side) may look like "attitude". This person may be "smiling" on the inside and very interested in the conversation.

**Big bursts of movement:** People in the more advanced stage of Huntington's often have difficulty their movement. For example, when getting up from a chair the muscles in their legs may use more force than needed to lift them off the seat giving the impression that they are leaping out of the chair. Another example could be with bathing: the person with Huntington's may try to gently lift their arm to help the caregiver wash them - instead they have a big burst of arm movement which is easy to mistake as 'aggression'. These uncontrolled bursts of movement may lead a caregiver to think that the person with Huntington's is kicking, hitting or resisting care. It is advisable to always be aware of these "big bursts." The caregiver should position both themselves and the person with Huntington's so that their safety won't be compromised. Over time, caregivers will learn more precisely how to anticipate these "big bursts".

**Smoking:** If the person with Huntington's smokes, they should be shown where they can smoke. If the person is a resident in a nursing home, it is important to establish what the rules are relating to smoking. If people require assistance and supervision whilst smoking, a daily routine and schedule should be set immediately and discussed with the team. It is important to remember that impulse control problems mean the person with Huntington's cannot wait, especially for a cigarette. They should be supervised closely for safety and smoking aids should be considered. It should be remembered that this person has suffered loss after loss: their job, driving, friends, their place in the family, their ability to live in their own home. Smoking often takes on a symbolic importance as "the only thing I've got left".

## Carers

When health and social care professionals are working with someone with Huntington's, they are also often working with their family carer. Remember:

- **They can be a great ally.**

No two people with Huntington's are the same; carers are often able to give great insight into the person. They should be asked about symptoms and strategies and about the history of the person. By the time someone is referred to Mental Health services there is often a very long history of concerning events. The snapshot received on assessment may be very different to the true picture. If the person with Huntington's won't engage a carer may be able to help. They may be able to facilitate them attending appointments or give feedback if they refuse to be there.

- **They need help too.**

Huntington's doesn't come with a manual. Everyone is different but there are common things that can help. The carer may value guidance on managing mental health symptoms, they may value guidance on what to do in a crisis – talk to them and listen to their concerns.


- **They are often very scared and very tired.**

A carer is likely to have been through so much - they may have dealt with or be dealing with other family members with Huntington's; they may have suffered extensive verbal and potentially physical abuse; they may feel like a failure; they may be at absolute breaking point; they are very likely to have been 'walking on eggshells' for a long time to try and prevent unwanted behaviour; they are very likely to have 'lost the person they love' and now taken on multiple roles (e.g. Mum, Dad and carer). All through this the person they used to depend on most in times of trouble is now the person they are caring for. It is helpful to talk to the carer to find out if they are coping or not. They should be involved if they want to be but their wishes should be respected if they would prefer to distance themselves.

## Further reading and resources

At the Huntington's Disease Association we have a team of Specialist Huntington's Disease Advisers (SHDAs) who cover England and Wales. We are able to give advice and guidance about Huntington's to families and professionals on a one-to-one basis or through formal training. We also have a team of Specialist Youth Workers who are able to work with children who live in a family where a parent has Huntington's and a Specialist Juvenile Huntington's Adviser who is available to support families and professionals in relation to Juvenile Huntington's disease.

Further information on all symptoms of Huntington's is available on our website  [www.hda.org.uk](http://www.hda.org.uk)

For information on research in Huntington's please visit HDBuzz which is a website that turns current research into readable information that is suitable for families  [www.hdbuzz.net](http://www.hdbuzz.net)





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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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phone **0151 331 5444**

**www.hda.org.uk**

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Registered Charity no. 296453  
A company limited by guarantee.  
Registered in England no 2021975

Design and print by Captiv8

Tel: 01892 611500 Web: [www.captiv8uk.co.uk](http://www.captiv8uk.co.uk)

Published April 2019 - Second edition

*Inspired by our community*