Huntington’s Disease Association

Mental illness and Mental Capacity in Huntington’s disease

A guide for mental health workers
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Foreword

Families affected by Huntington’s disease face many challenges. Perhaps the most difficult to cope with are the symptoms of mental illness that someone with HD may present with. Often families struggle to get access to the services that may help with this. This booklet was written following feedback from families, it is hoped that the information in it will help those working in the field of mental health support patients and families.

The Huntington’s Disease Association are indebted to Dr Hugh Rickards, Steve Hale and Doug Feery for their input and to Law 4U Solicitors for their financial contribution to funding this booklet.

Often small differences in treatment can have a huge impact upon a person’s quality of life, and in turn, that of the family as a whole. We hope this information supports you in caring for those affected by HD and their families.

Cath Stanley
CEO
Huntington’s Disease Association

Introduction

Huntington’s disease (HD) is a hereditary neurodegenerative disorder. Most people with HD develop the symptoms in their forties and fifties, although there may be subtle changes much earlier. About 5-10 % of patients 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 diagnosis is about 15 to 20 years, but some patients have lived 30 or 40 years with the disease.

The triad of symptoms of Huntington’s disease

Movement Disorder - the movement disorder is characterized both by the emergence of involuntary movements, or 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 patients become rigid and unable to initiate voluntary movements. Rigidity, spasticity and dystonia tend to emerge later in the course of HD, except in cases of childhood onset, in which they are often present from the beginning. Myoclonus, sudden brief jerks involving groups of muscles, is more common in juvenile-onset HD, where it may be mistaken for a seizure. Epilepsy is uncommon, though not unheard of, in adults with HD, but is said to be present in 30% of individuals with juvenile-onset HD.

Cognitive Disorder - the cognitive disorder is characterized initially by a loss of speed and flexibility. This may be seen first in complex tasks, when the patient 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 impairments in HD involve the so-called “executive functions” - abilities such as organization, 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 patients 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 persons with HD, both in the work place and at home.
Psychiatric Disorder - Psychiatric disturbances in Huntington’s disease are varied. The most common specific psychiatric disorder in HD is depression. Some patients suffer from conditions such as Major Depression, Bipolar Disorder, or Obsessive-Compulsive Disorder which are specific well-described syndromes, found in all sorts of patients.

Many, if not most people with HD also experience less well defined, non-specific changes in personality and mood, such as irritability, apathy, or disinhibition. Most of these psychiatric problems are believed to be related directly to the central nervous system injury caused by HD. Patients with HD 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 very devastating in HD, but they are also very treatable. Relieving a depression in someone with HD 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. Research is currently focussing on several aspects:

(i) a treatment that will delay or prevent the onset of symptoms;

(ii) for those people who are currently symptomatic, treatments that will slow down and hopefully reverse the symptoms;

(iii) research into effective treatment of symptoms.
Section 1 - Types of Mental Illness in Huntington’s disease

Mental illness is very common in Huntington’s disease. Even if the mental illness is caused by the Huntington’s disease it is still classified as mental illness. Sometimes people with HD 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 HD. 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 HD 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 HD.

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 HD on its own.

Other diagnoses
People with HD 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 HD but does occur. Sometimes it is difficult to tell what part the HD played in the causation of these illnesses but they can often be treated. Patients have a right to be seen and treated by mental health services.

Section 2 - Treating mental illness in HD

There is nothing about HD that prevents the treatment of any mental illness. As a general principle, patients with depression, anxiety and psychosis can be treated in the same way as people with those illnesses but no HD. However, there are a number of treatments for organic personality disorder which are effective but depend on the symptoms being treated.

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 / caregivers about the personality changes seen in HD can help them to change the environment, which, in turn, might lessen symptoms. Reducing excessive demands on the person with HD 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.
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 HD but is rarely complained of by people with HD and is difficult to treat. Often, managing the expectations of carers and spouses 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 HD and depression/anxiety commonly respond well to treatment.

**Psychosis**

Psychosis is rare in HD 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 HD and have a low threshold for testing for underlying causes, particularly chest and urinary infections and subdural haematoma. This are readily treated if diagnosed early.

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**Section 3 - The Mental Capacity Act and its guiding principles**

An individual with HD cannot be globally assessed as ‘not having capacity’. The MCA 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 HD cannot make complex decisions, this does not mean that he/she cannot make simple decisions;
- People with HD may lack capacity in specific areas. The mental incapacity may be permanent or temporary.
- The key principles in relation to HD

**Key Principle 1**

A person must be assumed to have capacity unless it is established that he lacks capacity. It cannot be decided that someone lacks capacity because they have HD 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 him to do so have been taken without success. The specific cognitive challenges that people with HD must be taken into account (see Section 5 assessing capacity and Section 10 cognitive impairment in HD).

**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 HD 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.).
**Section 4 - Assessing Capacity in HD**

The starting point for any test of capacity is an assumption that **the person with HD 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 individual 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 HD using the two-stage test of capacity**

People with HD 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 HD 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 HD might have specific cognitive problems which could impair this process.

1. **Perseverative thinking.** Many people with HD 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 HD 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 HD have a “mental disorder” as defined by the Mental Capacity Act?**

The vast majority of people with HD 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 HD who does not have a mental disorder (see section on mental disorders).

**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.

Important note 3: There can be circumstances where the individual with HD 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 untruths (e.g. can you describe how you get to the shop/how you prepare the meal).

**Section 5 - Best Interest**

The law requires that where decisions are being taken for/about an individual 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:

<table>
<thead>
<tr>
<th><strong>Key Principle 4</strong></th>
<th>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 his best interests.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Key Principle 5</strong></td>
<td>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.</td>
</tr>
</tbody>
</table>

A person trying to work out the best interests of a person 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**
- not 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**
- not 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. (See chapter 10 for more information about IMCAs.)

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.

Section 6 - Restraint

Restraint is defined in Section 6 of the Mental Capacity Act 2005 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 2005 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
- 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 an individual’s movements. A high level of control over an individual, 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.

Section 7 - Deprivation of Liberty Safeguards (DOLS)

The DOLS procedure aims to ‘safeguard’ the liberty of the an individual with HD 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 HD has to go through the DOLS procedure in many cases, once the individual 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 HD who struggles with ‘future visioning’ and may have negative memories from other family members) if the individual 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 HD 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 HD please contact 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 have an Independent Mental Capacity Advocate (IMCA) instructed to support and represent them during the assessment process. An IMCA is a specialist advocate trained to advise and represent individuals in relation to mental capacity issues.

Section 8 - Mental Capacity Act & Mental Health Act Pathways

All adults affected by HD, 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 1 above are followed at all times.

Mental Capacity Act Pathway
Overview of the deprivation of liberty safeguards process

Hospital or care homes managers identify those at risk of deprivation of liberty and request authorisation from supervisory body.

- Assessments commissioned by supervisory body, MCA instructed for anyone without representation.
- No refunds assessment.
- Age assessment.
- Mental health assessment.
- Mental capacity assessment.
- Best interests assessment.
- Eligibility assessment.

In urgent situations in hospital or care home can give urgent authorisation for seven days while obtaining a standard authorisation.

Best interest assessor recommends person to be appointed a representative.

Assessments supported authorisation.

Person or their representative applies to Court of Protection which has powers to terminate authorisation or vary conclusion.

Any assessment says no.

All assessments support authorisation.

Person or their representative requests review.

Managing Authority requests review because circumstances change.

Review.

Authorisation is given and person's representative appointed.

Authorisation implemented by managing authority.

Request for authorisation declined.

Any assessment says no.

Authorisation expires and managing authority request further authorisation.

Age assessment.

Mental health assessment.

Mental capacity assessment.

Best interests assessment.

Eligibility assessment.

No refunds assessment.

Any assessment says no.

All assessments support authorisation.

Person or their representative applies to Court of Protection which has powers to terminate authorisation or vary conclusion.

Review.
In addition to the above, consideration may also need to be given with regard to whether or not the Mental Health Act applies. This may consider if the individual may need to be assessed as to whether they ought to 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 HD are well recognised forms of mental disorder that subsequently present a form of risk to the individual, their health or safety, or to others.

The main Mental Health Pathways

![Mental Health Act Pathway Diagram]
Section 9 - Adult Safeguarding

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

The Social Care Institute for Excellence has identified that ‘Safeguarding’ encompasses six key concepts:
1. Empowerment
2. Protection
3. Prevention
4. Proportionate responses
5. Partnership
6. Accountability

Health and Social Care organisations play 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 investigating safeguarding concerns that are highlighted, but must do so lawfully, taking account of best interests, along with the rights of all those involved, in particular, the individual, the family and carers.

A generic overview of the safeguarding process is set out below, but note this does not highlight the need to involve the need to focus on best interest or the family!

Generic Flowchart - Implementing Safeguarding Adults Policy
Section 10 - The rights of Individuals, Families and carers

We have summarised a number of pathways that may be followed for an individual with HD 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 individual, 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, etc.

Often when these types of situations arise, there is agreement between all concerned, e.g. different family members, close friends of the person affected, 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.

Sadly however, a number of high profile cases 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. At the same time, and perhaps more seriously, in other cases where an individual had not lost capacity to make decisions, but the health and social care professionals involved had assumed that they lacked capacity as the decision being made in their view was an ‘unwise’ one, or that the individual needed to be ‘protected’ because of a perceived vulnerability.

Given this, it is crucial that there is clarity about the rights of:

- Individuals
- Families
- Carers

At the same time, it is also 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 individuals who may have difficulties that may arise from a progressive condition such as HD. The key rights are enshrined in the statutory principles referred to in section 1 above, namely:

1. A person must be assumed to have capacity unless it is established that they lack capacity.
2. A person is not to be treated as unable to make a decision unless all practicable steps to help him to do so have been taken without success.
3. A person is not to be treated as unable to make a decision merely because he makes an unwise decision.
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 his best interests.
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.

The rights of the family

What is clear from the above is that family members do have rights, most critically to be consulted about decisions that may need to be taken, but at the same time, family members need to understand that the individual with HD has rights, as highlighted above, and these must be respected.

One of the big difficulties for families is where they have cared for an individual for many years without any involvement by outside agencies from health or social care making the most if not all the decisions for their nearest and dearest. However, the family along with the outside agencies must observe and respect the rights of the individual, and where decisions need to be taken, then such needs to be done on the basis of best interest. Within this process, families do 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 may have a diagnosis of HD, is to take the opportunity to forward plan as set out previously. This way as a family member you can lawfully secure more decision making powers when the condition of your relative with HD 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 your 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 you object to the application being made. As explained in section 1, 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 individual cannot be admitted until the Nearest Relative has been displaced by the local authority obtaining an order displacing you as the 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 an individual appropriately

### Section 11 - Cognitive Impairment in HD

Cognitive impairment in HD 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 over looked. Cognition refers to the mental processes including attention, remembering, producing and understanding language, solving problems and making decisions. The word cognition is used to refer to mental functions and mental processes (thoughts).

Because different parts of the brain are responsible for different aspects of behaviour, it is significant that the frontal lobe is affected in HD. This is responsibility for organising, prioritising, controlling impulses, monitoring self awareness, motivation and finishing tasks, creative thinking and problem solving. This can lead to a variety of challenges when treating someone with HD. 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 unpicking 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 slower thinking, recall less fluent than recognition, difficulty with switching and maintain focus, difficulty in organising thoughts and actions, inability to wait for anything.

**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 HD 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 controlling the environment can be helpful.

**Recognition** is easier than recall means that a person's memory is not necessarily affected by HD. 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 you can help with this including distraction, setting boundaries by explaining this has already been discussed or setting a time amount for the length of time you will discuss it 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 HD. It means thoughts that used to be automatic now have to be conscious which makes it difficult for the person.
HD affects someone’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 HD.

**Section 12 - Forward Planning in Advanced Huntington’s Disease**

There can be little doubt that the significant life events we all may and will face – 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.

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

### Advance Decision to Refuse Treatment (ADRT)

An advance decision (sometimes referred to as 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.

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 (known as ‘life sustaining’ treatment). This includes treatments such as ventilation and CPR (cardio pulmonary resuscitation), which may be used if they cannot breathe or if their heart stops. If a person decides to refuse life-sustaining treatment in the future, their advance decision needs to be:

- written down
- signed by the person
- signed by a witness

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 advance decision, 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.

The Huntington’s Disease Association has created forms for making advance decisions available from Head Office.

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 individual’s situation, an advance decision gives the health and social care team clinical and legal instructions about the individual’s treatment choices. An advance decision will only be used if, at some time in the future, the individual is not able to make their own decisions about their treatment.

**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 2 types of lasting power of attorney (Individuals can choose to make 1 type or both)

- health and welfare property
- financial affairs

The Office of the Public Guardian (OPG - 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.

**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. You 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**

An individual 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 1 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 individual 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.

**Section 13 - JHD and Parental Responsibility**

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

The law imposes various age limits in respect of some pieces of the law, 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 bits of the law:

- Mental Capacity Act – 16 years and over
- Mental Health Act – 0 – until death (So no age limit)
- Deprivation of Liberty Safeguards – 18 years and over
So what about people under 16?
The general rule is that for those under 16, decisions for / 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 he is:
- married to the child’s mother at the time of birth
- listed on the birth certificate (after a certain date, depending on which part of the UK the child was born in)
- subsequently marries the mother
- applies to the Court for an order in respect of PR

In addition, there are other situations when PR can be obtained, for example:
- Where an individual is adopted
- Where an individual under 16 is taken in to the care of the local authority by means of an interim or full care order. In such situation, PR is shared with one or more parents who have PR
- 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 I am the young person and do not agree with my parent’s (or persons with PR) decision about me?
In this case, ‘competence’ of the young person would be looked at. What this means is that the age, understanding and ability of the young person to understand the issues at play 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. A famous case involved a 14 year female who wanted to be prescribed the contraceptive pill by her GP without her mother being made aware. Despite the concerns and objections of the mother after she found out that this had happened, the Court ruled against the mother’s objections.

**Section 14 - Day to day management of an individual with HD**

**Communication**
Due to the cognitive and motor difficulties it is necessary to give people with HD your time and patience when communicating. People with HD 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 HD occur because the muscles cannot move with the correct range of movement, speed, force or co ordination. One of 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 HD speaks eg 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 HD eg depression, personality changes such as irritability, lack of motivation can have an impact on communication. As HD 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.

Compliance: It is very difficult for people with HD to make changes to their routine and do ‘new things’. If you have received a referral for someone with HD there is a high chance that they will refuse to engage. This should not be seen as a reason to stop services; if anything it shows that they are even more in need. Options may be home visits (people are more likely to respond to this than being asked to come in to see you). Even on a home visit your entry may be refused, see if you can talk to family and establish when would be a good time to go, how they might best engage or find out if there are other professionals who are involved and they already have a relationship with then plan a joint visit. Remember if this person is in denial they aren’t going to understand the situation. Sometimes there may be no way of engaging initially, you may however be able to work with family and friends to support them in their caring role and work together to best support the individual.

Food and drink: Weight loss is an ongoing challenge to people with HD. People with HD may need more calories per day to maintain their body weight. People with HD tend to have ravenous appetites and are 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.

Beware of choking: As HD worsens, individuals develop a swallowing disorder. You may not be able to see that the individual is having difficulty swallowing, so you need to monitor and watch them whilst eating to prevent choking. People with HD 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.

Prevent falls: HD affects people’s mobility and balance. Pay close attention to walking and transfers to prevent falls.

See through the disguise: Weakness and changes in the tone of the facial muscles often contribute to an appearance of boredom. Difficulties maintaining a smile while listening or speaking may make a person with HD look unhappy, bored or uninterested. Weakness and changes in posture (such as leaning to one side) may look like “attitude”. Maybe you think the person just doesn’t like you. Don’t let this Huntington’s disguise fool you! This person may be “smiling” on the inside, very interested in what you’re saying to them and does like you. Don’t give up.

Big bursts of movement: People in the more advanced stage of HD often have difficulty controlling tone of voice and 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 HD may try to gently lift their arm to help you wash them - instead they have a big burst of arm movement which is easy to mistake this as ‘aggression’. These uncontrolled bursts of movement may lead a caregiver to think that the person with HD is kicking, hitting or resisting care. Always be aware of these “big bursts”. Position both the individual and yourself so that your safety will not be compromised. Over time, you will learn more precisely how to anticipate these “big bursts”.

Smoking: If the person with HD smokes, show them 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, set a daily routine and schedule immediately and discuss this routine with the team. Remember impulse control problems mean the person with HD cannot wait, especially for a cigarette. Supervise them closely for safety. Consider using smoking aids.

Remember: this person has suffered loss after loss: his job, driving, friends, his place in the family, his ability to live in his own home. Smoking often takes on a symbolic importance as “the only thing I’ve got left”.
Section 15 - Case Studies

**Robert – self neglect**

Robert is 61 years old; he was diagnosed with HD 8 years ago. He lives alone. In the early stages Robert was keen to engage with professionals who were there to support him (i.e. the specialist clinic).

Robert began to deteriorate physically and cognitively. He was in denial of the illness and, as such, didn’t think that there was anything wrong with him. He refused to get involved with health services. His adult children were concerned about the state he was living in (unwashed, underweight, carrying large amounts of money around, falling, and the house in an unhealthy condition).

Because of the relationship which had been built with the Regional Care Adviser (RCA) from the HDA, Robert was willing to engage with her (most of the time) and this allowed the RCA to facilitate a relationship with Robert and the Social Worker – by providing joint visits.

The police were called on two occasions when he wouldn’t open the door and was feared dead. He was referred to mental health services but was discharged because he refused to engage.

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**Bob – Difficult capacity assessments**

Bob is 45; he lives alone in his own house. With help from family he had coped OK and enjoyed the company of his dog, cats and rabbits. His house was well kept and his animals generally well cared for.

However, a neighbour expressed concerns about the welfare of the animals to the RSPCA; they visited and removed the animals, saying that the house was not fit to house them. Bob’s condition then deteriorated. He stopped washing himself, his clothes and his home. He refused help from relatives and became verbally and physically violent towards them.

The SW visited with the Regional Care Adviser from the HDA. They found a very dirty house; the carpet was wet with urine (from Bob who was occasionally incontinent and the new pets he had acquired), there was mouldy food, the toilet, bath and sink were full of faeces. Bob refused help.

Bob was targeted by a local gang who visited on the pretext of cleaning his house but were in fact stealing from him. They took him to the bank to get money out. Because of the theft the police were involved and the community officer visited on alternate days.
Bob attended clinic, he arrived looking clean, hair washed and all new clothes, as part of his assessment he was seen by a neuropsychologist who did a cognitive assessment which showed he had capacity to decide where he wanted to live.

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**Does Bob have a mental disorder?**

- **YES:** He could be detained under the MHA if he met the statutory criteria. There are grounds for admission based on Bob's health and safety e.g. the risks of the local gang who have targeted Bob.
- **YES:** Carry out a Mental Capacity Assessment to see if Bob has the capacity to decide where he lives (the cognitive test used is not sufficient). He would need to be asked to weigh up information including all of the perceived risks.
- **NO:** He is entitled to make unwise decision - guiding principles of MCA must be observed.

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**Deemed not to have capacity - Best Interest decision should be made**

Deemed to have capacity: the local team needs to continue to work with Bob. If he has been diagnosed as having a mental disorder this should include the CMHT.

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**Sheila - Moving into a care home**

Sheila has HD, she is 59 and she lives at home with her husband. She has become increasingly dependent on her husband over the years and he is no longer able to cope. Sheila has a carer who helps in the mornings and at bed time but if her husband is in the house she simply won’t engage with them, she will only take help from him. Sheila needs support with washing / dressing / toileting / eating but it is the constant demands that have taken the toll on her husband. He is emotionally drained.

Sheila has fallen twice when she has tried to get out of bed and she has hit out at her husband when he wouldn’t go to the shop to get more cigarettes.

Sheila has been deemed not to have capacity in relation to living at home (she was assessed on two different occasions, at home, by the social worker). A best interest meeting has taken place and the professionals and her family have agreed that she would be better off living in a care home. Sheila says that she will not move out, it is her home and she wants to stay there with her husband.

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**Sheila is deemed not to have capacity**

BI decision should be made, taking into account Sheila’s wishes as much as possible despite lack of capacity guiding principles of MCA must be observed.

**Important: What could be done for her to remain at home? Could this be on a trial basis?**

If not, this may result in a move: choosing a placement that understands HD, enabling Sheila to meet staff from the chosen home, enabling Sheila to go and see the home, enabling her to meet with other residents & look at a room.

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The MCA, providing you follow the rules, allows you to move the patient to a nursing home (including conveying the patient).

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Any placement would need to ensure that the placement of Sheila did not amount to a Deprivation of Liberty and appropriate safeguards were put in place.
John – placed on an acute ward.

John is 61 and has HD. He was admitted as a voluntary patient to a psychiatric ward after the risks of his self-neglect became too high. He has settled well on to the ward, he appears to enjoy having food there for him and has fitted in with the routine.

He wants to go home. John has been deemed not to have capacity to make the decision about going home and the professionals and his family are in agreement that this wouldn’t be safe nor in his best interests. John has been on the ward for 3 months and there are ongoing discussions about suitable placement and funding for such a placement so no progress has been made.

John was admitted voluntarily but now wants to go home. If not allowed to go home, under what authority is he being kept?

Consideration must be given immediately to whether or not the MHA or the MCA is to be applied

If MHA options would be S.2 assessment (28 days) S.3 treatment (initially up to 6 months) S.7 Guardianship (initially 6 months but renewable)

If under the MCA, does his placement at the Hospital amount to a Deprivation of Liberty? This may require urgent DOLS authorisation and/or consideration for an application to the Court of Protection

Consideration for John to be referred to an Advocate at the Hospital

If John had been admitted under the MCA he would have rights of appeal to Mental Health Review Tribunal for discharge. His nearest relative could also apply for discharge. If detailed under S.3 this would give an entitlement to after care meaning he may not have to pay care fees if he was placed outside Hospital.

The HMRT could be used as a vehicle to case manage a decision over placement where such was leading to John having to remain in hospital longer than was needed.

Paul – Violent Incidents

Paul is 46; he was diagnosed with HD 8 years ago. He has a history of violence with many convictions, episodes of self-harm and overdoses. He lives alone with no support. He drinks alcohol and smokes cannabis. Previously he managed at home with a care package and support from the local mental health team. However over a period of 4 years he had 5 care agencies, all withdrew due to violence and threats. Physical Disability Team felt they could not provide care due to level of violence although they did arrange and pay for him to have a daily meal at a local cafe. Sadly this broke down when he attacked the cafe owner and a customer.

Mental Health team discharged him; they believed he was choosing to live like this. He refused to attend the specialist HD clinic despite numerous appointments being made. He could not prepare a meal, his teeth were in appalling condition making eating difficult, he was losing weight, his home was squalid, he was vulnerable to financial abuse by others and could not successfully manage his money. Personal care was difficult for him and he was incontinent of urine.

At this point he was ringing the RCA 20 times a day wanting help with food, money and a variety of problems. He continued to be abusive to the public and the local police had numerous reports of his behaviour. The Physical Disability Team would only visit with a police escort and eventually declined to visit at all.

A multi-disciplinary meeting was held and funding eventually obtained for assessment in a specialist unit. The unit then felt they could not cope with his behaviour and would not accept him. Paul agreed to go into the
local mental health unit for assessment where they concluded that there were no signs of major mental illness and that he could control his behaviour. A cognitive assessment, while an inpatient, said he was functioning satisfactorily and he was discharged home with no support and a recommendation that future episodes be dealt with by the criminal justice system. His GP has now removed him from his list.

Paul attacked a member of the public and was arrested. After a period on remand he was released home, again with no support and has since attacked someone else. He is currently on remand again.

Court could be asked to transfer from remand to psychiatric Hospital

Invite the Court to Order Independent assessments

If found guilty, draw Court’s attention to disposal via a Hospital Order as oppose to a custodial sentence. Subject to the severity of the attack, the Court has a discretion to attach a Restriction order to a Hospital Order which means there would be ongoing oversight whilst the Restriction was in place by the MOJ. Even if given custodial sentence, if mental health condition deteriorated because of HD and sentence not expired, he could be transferred via MOJ warrant to a psychiatric Hospital.