Condition Insight Report (CIR)

Huntington's Disease

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Completed in collaboration with HDA

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Overview

What is the condition usually called / any abbreviations used?

Huntington's Disease (HD)

Some clinicians may still use the old outdated term 'Huntington's Chorea'.

Brief overview of the condition

Huntington's disease is an illness caused by a faulty gene in your DNA. If you have Huntington's, it affects your body's nervous system.

Huntington's can cause changes with movement, learning, thinking and emotions. Once symptoms begin, the disease gradually progresses, so living with it means having to adapt to change, taking one day at a time.

Juvenile Huntington's disease (JHD). Manifests under the age of 20 and has slightly different symptoms such as rigidity and an increased propensity for epilepsy. Individuals with a diagnosis of JHD can live into adulthood.

What is the generally preferred term for someone with this condition?

An individual living with HD.

Presenting Symptoms

In the early stages (before a motor diagnosis is made) the individual's cognitive skills may be affected and could include restrictions with:

- Ability to multi-task, processing information and speed of processing, inflexible thinking, inability to self-monitor, impulsive behaviour and short term memory. Alongside apathy.

Some research shows that the brain can begin deteriorating up to 20 years before motor diagnosis, so cognitive/psychiatric symptoms may appear well before motor symptoms. People with HD might need a lot of help through prompting and supervision/assessing risks early on, much more than their physical symptoms might suggest. It is very important to take this into account.

At the stage that the person is diagnosed they will have marked motor symptoms (movement disorder) Symptoms will include but not limited to:

- Uncontrollable involuntary movements - flailing arms and legs (chorea)
- Difficulty with voluntary movements (e.g. opening/shutting a door, body control when sitting down)
- Slowness of movement (bradykinesia)
- Difficulty with walking/mobility (partly due to spatial awareness)
- Inability to recognise personal space so could come closer than others would or bump into furniture
- Reduced dexterity and grip
- Fatigue
- Reduced executive functioning
- Weight loss
- Reduced swallow
- Slurred speech
- Visual perception and saccades is an issue
- Incontinence
- Dizziness

It is important to note that many people with HD suffer from *anosognosia* a complete unawareness of their symptoms - a lack of Insight. This prevents them from seeking help and can make the provision of multidisciplinary care a complex challenge. This might also mean that people with HD might not have the services or evidence in place to fully demonstrate how their condition is affecting them.

A small percentage of individuals will have further psychological symptoms such as delusions and hallucinations.

Fluctuations

There is degeneration in function over time but people do often have worse days generally following over exertion. Individuals cope much better with a routine. If they have to do something out of the ordinary this can cause a huge amount of anxiety in advance, a very challenging day, and exhaustion for days after.

There is no single set pattern to the degeneration process of HD; it can be uncertain with the length of time with prognosis being unpredictable (being dependent upon other health issues). Each individual can degenerate differently and at different rates.

Think about exploring things like:

- **Cognitive function**
  How consistent is their presentation, how do they manage their memory deterioration, are they orientated, can they plan tasks that they are used to? How long does it take to do a task? *Are they giving a true reflection of their ability? – consider exploring this with a companion.*
- **Consider insight**
  Have they noticed the changes in their ability and do they recognise what is happening to their body?
- **How has the condition been over the past 12 months?**
  What changes have they seen and over what period of time? Have they experienced any deterioration? If so, what has this meant for their functional ability now compared to previously?
- **What level of pain/fatigue do they experience?**
  Can they quantify this? Do they use a scale and can they describe it? Can they share what a ‘typical’ day might be for them? What are their triggers? Whilst exertion is a main trigger what amount of exertion is enough to cause further limitation? How are triggers managed?

Reliability

What specific areas should be covered to ensure a complete, reflective report?

SAFETY	TIMELY	ACCEPTABLE STANDARD	REPEATEDLY
Do they have any symptoms which could cause a safety consideration?	For any activities where restriction is reported how long does it take them to complete these activities? Has how long it takes them changed over time?	How have they adapted to completing tasks over time – is this different to what might be considered ‘normal’?	Are they able to repeat a task as often as required? Is this the same every day?
Due to the varied symptoms which may impact safety, this is a critical area which needs to be explored. Symptoms such as uncontrollable involuntary movements and cognitive deficit are clinically likely to impact someone’s ability to complete an activity safely. Furthermore, if they lack insight, this may lead to them under reporting their functional restrictions. Due to lack of insight or cognitive deficit, some people may think they are completing activities safely, but in fact they are not.	Activities and jobs that require fine motor control of the hands may no longer be possible for the individual to undertake. It can take up to 3 hours for the HD individual to prepare for the day ahead. Each action: from waking, getting out of bed, washing, dressing, and eating, can have long delay. Physical and mechanical difficulties, short-term memory issues and other symptoms can cause unexplained stop - start actions.	For most people apathy is an issue so without support many would simply not complete tasks or make their appointments without the support of others. However as denial of HD, or little/no insight is very common, they may be unaware of any symptoms at all and may deny the diagnosis and support required. Probing effectively to determine how tasks are managed is critical.	Due to the degeneration that occurs over time and the variation between individuals this needs to be explored for all relevant activities. You should try to explore how someone may be before, during and after activities where a restriction is present to determine fully whether the task is repeatable in the same way as often as required throughout the day, especially with elements like fatigue affecting their presentation.

Sensitivities

What areas might they find difficult to mention or perhaps understate the impact of?

It is important to remember that most of the symptoms of HD are hidden because they are cognitive/behavioural and many have poor insight so struggle to provide the information themselves.

Other areas that might be difficult are discussing:

- Their physical symptoms and changes to their overall function or independence
- How the disease will progress
- The behaviour they experience from others who do not acknowledge or understand their condition, especially family/friends
- Consider if you are exploring cognitive decline with a companion, that this may cause tension with the claimant and should be managed professionally.

Watch a video of Matt talking about HD **HERE**

Customer Care

How is it best to ask about any sensitive topics and what are the common courtesies?

In general

- Aim to have very little distraction e.g. background noise, shuffling papers etc.
- People will feel very stressed and anxious about an assessment whether at a centre or over the telephone. Give them time. Let them have short breaks if they seem agitated or if anxiety is increasing
- It is really important to ask appropriately short, probing questions. A series of itemised routine questions could gain good responses
- Always offer the person who accompanies the individual with HD the chance to speak. If the person with HD is unaware of their symptoms, their partner/carers etc will be a much more reliable resource. This wherever possible, should be done out of earshot of the claimant, but with consent, as it could result in a backlash for the accompanying person during or following the assessment

During face to face interactions

- Keep things simple and sit directly facing the HD individual
- Ask a basic closed question, that requires a yes/no, or a single word answer, clearly, then wait for a response. Your speech doesn’t need to be slowed down. A person with HD can usually understand and answer well but it takes time for them to find the answer (the words to construct the reply). Don’t interrupt them when they are formulating their answer - otherwise they will have to rethink and start all over again
- If someone is at the stage where their speech is difficult to understand then give them time. Ask them to repeat the reply or words not understood and use the skills of the person accompanying them
- The individual with HD may not wish to dwell on commenting on their condition, and may have got used to underemphasizing the effect it has on them as a coping strategy, in order to get through each day

Functional Impact

A brief summary of the functional impact those living with this condition may experience

Activity 1: Preparing food

In the very early stages (before a motor diagnosis is made) the individual's cognitive skills will be affected. This affects the ability to multi-task, speed of processing, inflexible thinking, inability to self-monitor and impulsive behaviour and short-term memory is breached. Apathy affects the HD person's ability to be motivated for tasks. Poor motor control will progress until the individual is likely to require full support with this task. They may have experienced injury whilst they have limited insight.

Remember in PIP…

Can someone prepare a meal reliably? Someone's physical ability to complete an activity is not the only area to be considered here. If there is evidence of cognitive deficit, are they safe cooking? Can they tell if food is cooked? Is the food safe to eat? Are they safe around knives and flames? Furthermore, if there is a lack of motivation, do they require prompting to cook? Are they cooking the majority of days? Do they require support or supervision? If there are issues with motor skills, can they safely cut food or hold pans? Do they require physical assistance?

Activity 2: Taking nutrition

Some people with HD need to maintain a 5,000 calories a day food input. They may be starting to experience issues with swallowing. Weight loss becomes an issue at this stage.

Remember in PIP…

You need to have established the individual's ability to be nourished, either by cutting food into pieces, conveying it to the mouth and chewing and swallowing; or through the use of therapeutic sources. Are they at risk of choking? Do they require soft or blended food? If there is weight loss secondary to their condition, do they require prompting to eat regularly? Is this prompting throughout the duration of the meal?

Activity 3: Managing therapy and monitoring a health condition

Some individuals with HD may have a lack of insight, and consequently management of their condition may be poor.

They may require drug therapies, speech and language therapy, occupational therapy and/or physiotherapy. Increased loss of control on movements and cognitive changes will mean many require support.

Remember in PIP...

If someone lacks insight, you must establish how they are managing their treatment. Do they require prompting or support to administer their medication? Do they have poor compliance?
Furthermore, if they receive therapy in the home environment, do they require assistance with this? How long does this assistance take?

Functional Impact

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Activity 4: Washing and bathing

Speed of processing, inflexible thinking, inability to self-monitor, impulsive behaviour and short term memory may mean an individual is prompted to complete personal care.

Apathy affects the HD person's ability to be motivated for tasks. Poor motor control will progress until the individual is likely to require full support with this task. The HD individual's home is likely to have corridor rails and grab bars in the wet room or bathroom. The HD individual will, most likely, use a shower stool or chair as standing and turning in the shower is a complex manoeuvre. Equipment may be refused by some but lack of it is not an indication of ability.

Remember in PIP…

You must not only consider physical ability to complete the activity, but also recognition of the requirement to wash and motivation. Do they require prompting to wash regularly? In the absence of prompting, how often would they wash? If they have a physical restriction, how are they managing to wash their entire body? Can they physically get in and out of an unadapted bath? How long can they stand for? Do they require assistance to wash their body? If so, is this their entire body? How long would it take them to wash unassisted? Would this be acceptable when considering the extent of their symptoms?

Activity 5: Managing toileting needs and incontinence

Once the physical aspects of the disease are evident, most people at this stage would be struggling with toileting and may be incontinent. Fine motor skills become a constant problem. The HD individual may also have a specially installed toilet, maybe with guard rails and intricate personal cleaning facilities. Equipment may be refused by some but lack of it is not an indication of ability. For some individuals with HD, the message from the bladder to the brain is so slow that it only gives a very short time to get to the toilet and this, alongside mobility problems, can cause incontinence.

Remember in PIP…

You must explore not only someone's ability to get on and off the toilet, but also to reliably clean themselves. This will be a sensitive area to explore, so should be probed delicately. Can they reliably clean themselves after going to the toilet? If they have aids, can they reliably use them or do they require assistance? If they report incontinence, can they self manage? Is this of both bladder and bowel?

Activity 6: Dressing and undressing

Apathy affects the HD person's ability to dress and undress. Individuals will have difficulty with fine motor movements, such as using buttons or zips, due to a lack of control and strength in their hands and fingers.

Remember in PIP…

How do they manage to dress themselves? Are they capable of selecting appropriate clothing? Do they require prompting to do this? If they can dress, how long does it take? Can they manage fastenings, buttons and laces? How do they feel after? Do they require assistance or use any aids to dress?

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Activity 7: Communicating verbally

Speech can be affected due to the effects on the muscle functions, which can make it difficult to control volume and clarity of speech which can lead to others misunderstanding what they are trying to express.

Remember in PIP...

The scope of the activity is to be able to both express **and** understand. It is not enough that someone can hear what another person is saying. Can they process it and respond appropriately and in a timely manner? How do they manage with communicating with others? Do they require support to do so?

Activity 8: Reading and understanding signs and symbols

Visual perception and saccades is an issue (difficulty with reading, seeing, scanning and avoiding obstacles etc). With any cognitive changes, this can affect the ability to process and understand written information and in some cases support is required.

Remember in PIP…

Simply being able to read information is not adequate to manage this activity. The information someone is reading must be **understood**. What level of information can they understand? Although retention of information is not considered, it is clinically reasonable that someone will have difficulties processing, and this should absolutely be explored during the assessment.

Activity 9: Engaging with others face to face

Anxiety, agitation, apathy and other cognitive changes as well as affected speech can affect an individual's ability to engage socially.

Remember in PIP…

If there are cognitive changes reported, you must explore their ability to safely engage with others. Are they vulnerable? Are they at risk when engaging? Do they require support? If they have anxiety, who can they engage with regularly? How do they manage engaging with unfamiliar people? Do they require support from a specific individual to do so? **REMEMBER: Just because someone reports they can mix with others, this does not mean they are doing so safely.**

Functional Impact

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Activity 10: Budgeting

In the very early stages the individual's cognitive skills will be affected. This affects the ability to multi-task, speed of processing, inflexible thinking, inability to self-monitor, impulsive behaviour and short term memory is breached. Apathy affects the HD person's ability to deal with budgeting. Bills and mail may not be opened due to apathy.

Remember in PIP…

If someone has a cognitive deficit it may be that they are unable to prioritise finances. Could they reliably manage a household budget? Do they require support? How would they cope with an unexpected bill? Can they manage future financial planning? Also, could they understand change expected in a shop?

Lack of motivation and anxiety alongside impulsivity must also be considered here.

Activity 11: Planning and following a journey

In the very early stages (before a motor diagnosis is made) the individual's cognitive skills will be affected. This affects the ability to multi-task, speed of processing, inflexible thinking, inability to self-monitor, impulsive behaviour and short term memory is breached. Spatial awareness related to calculation (unable to maintain bearings both domestically and geographically). Crossing roads can become a major problem. People are often resistant to change/advice etc. They don't 'realise', they aren't 'aware', can't 'predict', having difficulty with organising and can't easily analyse situations. There is no mental agility (acute difficulty organising daily activities, processing sequential activities, etc).

Remember in PIP…

You must explore safety in this activity. If someone is completing journeys, are they safe? Do they have road safety? Are they impulsive around cars? Also could they maintain concentration when out of the home? Are they vulnerable? Could they manage a diversion or plan and follow a new route? How would they cope on public transport? What would they do if they got lost? Have there been any incidents when out of the home? How do they cope with distractions?

Activity 12: Moving around

People may fall over for no obvious reason, but not to a degree that medical intervention is needed. Gait and balance issues mean that many HD individuals walk with a staggered movement. This makes it difficult to walk uphill and downhill. Stepping up or down from a kerb requires a lot of concentration. If people are trying to go from sitting to standing, they may 'jump up' and stagger towards an anchor point e.g. a wall to stop. Many HD individuals have to use a wheelchair for part of their day - or all of the time.

Just because HD is not a musculoskeletal condition, does not mean it will not substantially impact mobility. The nature of the condition is such that it will limit their ability to reliably move around.

Remember in PIP...

You must explore STAR when asking about someone's ability to mobilise. It is not sufficient to simply indicate a distance or time someone can walk for.

Firstly, establish when someone is walking, do they require an aid? If so, what aid? Was this prescribed or self purchased? How often are they having falls? How do they manage this? What is their gait?

How far can they walk and how long does this take? How does walking make them feel? Does breathlessness/fatigue increase each time they walk? When they walk a second time, does pace reduce or distance reduce? If they experience fatigue, how do they feel later in the day? How many times could they repeat the distance they walk? How long does it take to recover from mobilising? How do they cope with uneven surfaces?

Lived examples are also extremely beneficial. How do they manage up and down the stairs? How do they manage with chores? How do they manage their shopping? How many aisles can they walk and how long does it take? If they go to their GP surgery or hospital, how far is it from the car? How long does it take them to get to the required department and how far is this?

Additional reading or other resources

EXTERNAL

- www.hda.org.uk
- https://www.hda.org.uk/get-involved/campaiging/huntingtons-disease-awareness-month-2023/cognitive-impairment/
- https://www.hda.org.uk/huntingtons-disease/living-with-huntingtons-disease
- For further information including medications see NHS Conditions and Treatments: https://www.nhs.uk/conditions/huntingtons-disease/
- https://www.youtube.com/channel/UCiO73l6SP8_x6Rci_ZX3ZUA

INTERNAL

- Desktop Aid – Fatigue, Activity 12, Washing and Bathing, Dressing and Undressing with or without an Aid, CSE

Version control

Version	Date	Signed off by	Comments
1.0	03.06.2020	Dr Shah Faisal and HDA	New re-banded document
1.1	03.02.2023	Rebecca Sparks	General review completed to ensure up to date guidance is being utilised. Amendments made to 'Remember in PIP' areas across activities
1.2	17.04.2023	Rebecca Sparks and HAD	Stakeholder document review with comments added to presenting symptoms STAR, fluctuations and customer care.
1.3	27.09.2023	Jade Mayfield and HDA	Stakeholder document review with added link for cognitive restrictions