Huntington's Disease Association

NHS Continuing Healthcare (CHC) assessments for people with Huntington's disease

Contents

About Huntington's disease

Huntington's disease is a rare, genetic, neurodegenerative disorder and there is no cure. Around 8,000 people in the UK have the condition. It is caused by a complex genetic mutation that leads to widespread and irreversible brain damage, making it difficult to develop effective treatments. Every child conceived naturally to a parent who has the faulty Huntington gene has a 50% chance of inheriting it. If a person is 18 or over, they can take a genetic test to find out if they have the faulty gene.

Symptoms can start at any age, but they commonly appear between 30 and 50. Changes in personality and cognitive changes, such as social understanding and organisational ability, can start up to 15 years before the clinical diagnosis, which is based on changes to movement. For those under the age of 18, the assessment and the decision to diagnose can be challenging.

CHC Assessors are advised to liaise with the person's neurology/ Huntington's disease service in charge of the person's care for a better understanding of their current needs and treatment. Many of the symptoms between adult and juvenile onset Huntington's disease are similar. However, in people with juvenile onset Huntington's disease, rigidity of the body is more common and the rate of decline can be more rapid.

Symptoms of Huntington's disease can include:

- Involuntary movement (movements may happen that you cannot control, making everyday activity more difficult)
- Cognitive (difficulties in thinking and processing information)
- Mental health (changes in behaviour and personality)

Adult-onset Huntington's disease is usually fatal after a period of up to 20 years. In the later stages of the disease, people will find communication and daily activities increasingly difficult and need full-time nursing care.

Purpose of this guide

This guide is designed to support CHC Assessors to understand Huntington's disease in the context of the CHC domains, to ensure that people living with this rare, neurodegenerative condition have a fair and accurate assessment of their needs.

In order to collate information for the assessment, CHC Assessors should be aware that whilst specialist Huntington's disease services do exist in England and Wales, their commissioning and resourcing are not all equal. This means they may not see Huntington's disease patients as often as they would like or need, and have to share care across geographies and teams. CHC Assessors are advised to liaise with the person's neurology/Huntington's disease service to understand their pathways and resources.

CHC Assessors should be aware that around 4 in 10 people with Huntington's disease (38%) live alone , which can sometimes be a consequence of personality changes leading to family estrangement. In combination with any cognitive change, which can include a significant lack of insight, it may be difficult to gain a true picture of that person's day-to-day function. We recommend that CHC Assessors seek out a proxy report in these cases. If unsure, assessors can contact the Huntington's Disease Association or the person's neurology/Huntington's disease team for advice. For the same reason, it is also good practice to do this even when support networks are present.

Contributors

Alex Fisher

Alex is a Senior Occupational Therapist in Neuropsychiatry at Birmingham & Solihull Mental Health Foundation Trust, who specialises in supporting people living with Huntington's disease. She is also the Huntington's Disease Module Lead for Neuropsychiatry, at the University of Birmingham.

Lilian Wiles

Lilian Wiles is a Registered Nurse. Lilian's current role is as an All Age Continuing Care Case Manager – Appeals, at NHS Birmingham & Solihull Integrated Care Board.

Lilian qualified as a registered nurse in 1988. Her career has spanned primary, community and secondary NHS care settings, in addition to palliative care, hospice and private health care roles.

Lilian is also a Fellow at the Health Foundation and completed a PGC in Leading Quality Improvement in Healthcare as part of the Generation Q project.

Huntington's disease in the context of the care domains

1. Breathing

People with Huntington's disease will experience respiratory decline and struggle to cough and clear airway secretions effectively. Silent aspiration of secretions adds to the risk of pneumonia. Pneumonia is one of the most common secondary causes of death in Huntington's disease. Involuntary movements caused by the disease will also affect the muscles involved in breathing. People with Huntington's disease, particularly in the mid-to-late stages, may not be able to 'feel' the challenges associated with any breathing difficulties, due to changes in their ability to process information.

Due to communication challenges, they can also struggle to tell people they are struggling to breathe. They may not understand the changes and will struggle to engage in or tolerate any intervention. People with Huntington's disease will develop dysphagia. Along with the respiratory decline, this can exacerbate the risk of food/fluid entering the lungs. Dysphagia results from impaired voluntary control of the mouth and tongue, impaired respiratory control due to chorea and impaired judgment; resulting in eating too rapidly, or taking overly large bites of food and gulps of liquid.

Questions To Consider

Breathlessness

- How does the person describe their own feelings of being breathless or unable to breathe? Are they able to describe them?
- Has the person seen a professional about their breathing? If yes, have they been able to engage with that professional and been given any advice? Are they reliant on a trusted other, such as a family member, to support compliance with any advice or intervention?
- Is the exertion of daily living activities leading to breathlessness? Does it take longer to complete care tasks, because the person becomes breathless and needs to rest/recover? Do breathing difficulties affect their ability to participate in care and/or complete care tasks independently?

Anxiety, distress and respiratory function

- Is the person experiencing shortness of breath or breathing problems associated with anxiety or extreme distress? Have they received any support for this? Are they able to engage with this?

Other respiratory conditions

- Does the person have any other respiratory condition(s) requiring treatment with medication? N.B. This could include medication that is prescribed regularly, or as required e.g. reliever medication for asthma, or oxygen therapy.

Respiratory interventions

- Does the person have increased oral secretions?

Questions To Consider (cont.)

- Do they need support to maintain open airways, such as suctioning? Does the person require a respiratory aid, such as a cough assist machine, due to a reduced ability to cough? Are they able to engage with this and tolerate it?
- Is the person experiencing chest infections or coughing when eating, which may indicate problems with swallowing? If the person is experiencing chest infections, how frequently are these occuring? How long do they last? And how are they being treated?
- Is medication or other therapies prescribed to optimise breathing and/or reduce the impact/severity of breathlessness symptoms? If yes, how often is this kind of intervention required in a 24-hour period and who provides it?

Respiratory function and involuntary movement

- Does uncontrolled chorea or dystonia affect the airway and posture?

2. Nutrition – Food and Drink

As Huntington's disease progresses and more symptoms appear, people often find it more difficult to maintain their weight and to prevent significant, unintentional weight loss. The person will often burn a high number of calories due to chorea. Unintentional weight loss is also part of the disease pathology and presents challenges in consuming enough calories in a safe and dignified manner. People may also be neurologically apathetic and not have the drive to eat or to 'feel' they need to eat. The person can struggle to get food or drink to their mouth, prepare food or work out new ways of eating. They may not comprehend the need to eat more.

Swallowing problems, also known as dysphagia, can lead to difficulties whilst eating and drinking due to an increased risk of choking. A PEG (percutaneous endoscopic gastrostomy) tube allows food, fluid and medicines to be received directly into the stomach. This can be introduced if it is too difficult or unsafe to do this by mouth, due to the effort and risks associated with swallowing. A PEG does not prevent aspiration of secretions such as saliva. Some people may have an advance decision to not have a PEG, others may refuse to have one or others may be too frail to receive one. Any decision should be on a case-by-case basis and consider the person's ability to understand the issues.

Questions To Consider

Professional guidance, intervention and engagement

- Has the person seen a professional to discuss concerns about eating and drinking? E.g. speech and language therapist or dietician. Are they able to engage with any advice or intervention? If not, why are they unable to engage? Can the person communicate in a reliable way if they are hungry or thirsty?
- Are there any pre-existing conditions, habits, preferences or other cultural or environmental/ contextual barriers to maintaining optimum nutrition and hydration e.g. if a person has anorexia or a tendency to limit food intake?
- How frequently are there difficulties encountered with the delivery of prescribed fluids and feeds?
- Have there been any assessments from a specialist occupational therapist to help identify interventions to promote independence with eating and drinking?
- How are fluids delivered? E.g. adapted beakers, straws, etc.
- What would a usual portion size look like for any given meal (think in terms of bowl or plate size)? How is it eaten? E.g. teaspoon, dessert spoon, finger foods. Is the person able to use normal cutlery, including for cutting and spreading? How long does it take the person to consume a usual portion of food or fluids?

Weight loss, calorie need

- Is the person a healthy weight? Has their body weight, BMI and nutritional status changed over the last 3-6 months? If it's not possible to weigh or monitor body weight regularly, what other measurements are being used to assess nutritional status? E.g. upper arm circumference measurements, handgrip, changes in clothing sizes, blood tests.
- What are the person's calorific needs and are there any challenges in meeting this need?
- Is there a prescribed target in relation to calorie intake or fluid intake in 24 hours? If yes, what is the target? Who is monitoring this and how is it being reported back to the prescriber?
- Does the person require supplementary feeding or enriched diet? If yes, does this require monitoring from a dietician? Do they have insight into the need to have an enriched diet? If not, what risks does this present?

Choking, vomiting, aspiration

- Do they have choking episodes? E.g. poor coordination leading to choking on liquids and solid food.
- Is there anxiety around eating (e.g. fear of choking)? If yes, what is the impact of this?
- Are there any specific requirements for preparation prior to or after offering any oral intake, to reduce the risk of choking, aspiration or regurgitation/ vomiting and silent aspiration?

Questions To Consider (cont.)

- What, if any, escalations/ interventions have been required to reduce the incidence of choking, aspiration, regurgitation/ vomiting and how frequently have they been needed/how has this changed in the last 3-6 months?

Fatigue, (cognitive) attention

- Does the person struggle to finish a meal? E.g. they are easily distracted by TV and other people having a conversation and/or they are too tired.

Behaviour - perseveration

- Are there other challenges associated with behaviour/ perseveration, which is making it harder to have an adequate diet? N.B. Sometimes people with Huntington's can get 'stuck' on certain ideas and find it difficult to move on. This is known as perseveration. This may also be apparent when someone with Huntington's becomes obsessed with a particular item and buys it in excess.

Eating, drinking and support

- Is there a requirement for someone to prepare, serve and oversee a modified consistency diet (or a fortified one) and/or fluids - what is the prescription for this and how is it monitored for safety/efficacy?

PEG

- Does the person require a PEG tube? If yes, have there been complications? If they are not using a PEG tube, has it been suggested they have one but they have decided against it?
- If an enteral feeding regime is in place, e.g. via PEG or similar, who is managing and maintaining this regime? What additional knowledge, skills and experience are required to maintain this safely and ensure it is delivered in a timely way?
- If the person has a PEG, is this for all of their nutritional needs, or do they have a combination of enteral and oral feeding? N.B. PEG feeding may be to supplement oral intake to achieve nutritional goals to maintain healthy weight and energy levels. When the majority of nutrition is administered through a PEG, "taster feeding" small amounts of food, can be important for pleasure and maintaining oral skills.

Nutrition, hydration and involuntary movement/ coordination

- How do the unpredictable movements and changes in posture/balance affect the person's ability to eat and drink safely and optimally?

Oral health

- Has the person experienced a deterioration in their oral health? E.g. problems with mouth/gum infections that affect their ability to eat and drink comfortably. Can the person attend to their oral health (e.g. brush teeth) independently? What are the barriers to this?

3. Continence

Huntington's disease affects the processing of any signals related to bladder and bowel function. Incontinence is a challenge for many people with Huntington's disease. In the mid to advanced stages of Huntington's disease, people usually become firstly incontinent of urine and latterly faeces. Regular pads may be ineffective, for example, if a person has chorea (involuntary, irregular or unpredictable muscle movements). Some people require catheterisation.

People who are still continent often experience urinary frequency and urgency, which can necessitate frequent visits to the toilet. Altered bowel habit, such as constipation, can become problematic and have an impact on mood, particularly through increasing irritability. Cognitive impairment may prevent the person from adopting any suggested intervention. Self-monitoring becomes increasingly difficult and this includes remembering to change soiled incontinence products and attendance to hygiene, resulting in cross-infection and skin problems. Symptoms such as perseveration may cause the individual to become hyper-focused on toilet habits.

Questions To Consider

- Is the person incontinent? If yes, is it bowel and/or bladder, and is it day and/or night?
- Is the person known to the community continence services and had their individual continence needs assessed in line with NICE guidelines? N.B. Assessment should also take into account any neurobehavioural characteristics.
- Is the person concordant/able to cooperate and accept help with continence care?
- Is the person struggling to get to the toilet on time due to the impact of physical symptoms and problems with mobility? E.g. involuntary muscle contractions (dystonia) and unusually slow movements (bradykinesia). Do they need help to get to the toilet? Does the person need support to transfer on and off the toilet?
- Can the person cleanse themselves and the environment after a visit to the toilet? Do they appear soiled in family and public spaces? Are they aware of this?
- Has the person been referred to a specialist occupational therapist to look at products which might enable a safer, more comfortable and more dignified toileting environment? (I.e. washer/dryer toilet, reinforced or padded seating to minimise breakage and injury.)

Questions To Consider (cont.)

- Are involuntary, irregular or unpredictable muscle movements making continence care more difficult? E.g. pads moving around and becoming torn, catheters leaking, bedding/bed pads needing to be changed.
- Due to perseveration (repetitive ideas), is the person asking to go to the toilet frequently? Has any treatment been trialled for this? Does the person have an aversion to going to the toilet – perhaps having developed unusual thoughts about the experience? Might they only go to the toilet in specific places?
- Are there challenges to supporting the person's continence needs in a 24-hour period? E.g. reluctance to accept personal care following incontinence, refusal to use incontinence aids.
- Does the person have a history of urinary tract infections (UTIs)? If yes, has this affected continence?
- What does a normal bowel habit look like for the person and how is this maintained?
- Have there been any hospital admissions or referrals to the out-of-hours services because of urinary or bowel symptoms?
- What additional medications/ interventions are prescribed to promote bladder function (including urinary catheters, urostomy, or use of urinary sheaths) and/or prophylactic antibiotic therapy? Is the person cognitively able to follow this advice or physically able to manage any advice?
- What additional medications/ interventions are prescribed to promote a regular bowel habit? E.g. laxatives (type and frequency) and/ or stoma. Is the person cognitively able to follow this advice or physically able to manage any advice?

4. Skin (including tissue viability)

The constant, uncontrollable movements (chorea) people with Huntington's disease experience are exhausting. Friction, combined with inherent weight loss, can result in skin breakdown. An inability to regulate temperature may cause increased perspiration and make this worse. Frequent repositioning and specialised seating/beds can help, although finding suitable seating and beds can be challenging when a person has involuntary muscle movements. As a person's mobility deteriorates, they will experience falls and increased tissue trauma. Cognitive impairment and hyposensitivity to pain will influence their ability to seek support, monitor and engage in treatment, leading to non-deliberate neglect of skin and skin damage.

Questions To Consider

- Is the person developing marked or broken skin due to friction? Are involuntary, irregular or unpredictable muscle movements leading to a risk of skin breakdown, which requires monitoring? Are they aware of the problem?
- Is the person under the care of district nurses/GPs for help with wound or skin trauma care? Has the person received support from a tissue viability nurse? Are they able to engage in assessment, support and treatment? If not, what are the barriers?
- Does the person require pressure care/ position changes/equipment to reduce the risk of pressure damage? What equipment is in place to reduce the risk of avoidable harm and deterioration in skin integrity? Who is responsible for managing and maintaining this to ensure it remains appropriate? Are there any prescriptions for any pieces of specialist equipment (including chairs, pressure cushions, mattresses, beds, or footwear etc.)?
- Does the person experience perseveration (repetitive ideas) or delusions (due to psychosis) related to their skin? E.g. around itching. If yes, what treatment is being trialled? Has it been effective?
- Are there any other underlying skin conditions, skin lesions, allergies or health conditions that have the potential to undermine skin integrity? If yes, how are these skin issues usually managed and is it a recurrent problem?
- Is the skin intact? If not, what are the details of the wound/ lesion characteristics and is there a management plan?
- What is the Waterlow or other risk assessment score for this person and how has this changed in the last 3-6 months?
- What, if any, treatments are prescribed (this might be solely equipment) to promote and maintain skin integrity and how often is it used?
- Who is providing care and management of hair, scalp, beard, mouth/dental hygiene, fingernails, feet and toenails? Are there any problems with any of these?

5. Mobility

Balance, coordination and walking problems are common for people with Huntington's disease. A high proportion will have recurrent falls and these will increase with advancing illness until they transition to immobility. Skin trauma and fractures are common as is brain injury, such as subdural haematoma.

Regular multi-disciplinary team assessments are required to assess risks related to mobility and identify how to work with the person and their caregivers to minimise risks. Mobility aids may be useful if the person is cognitively and physically able to operate them, but often can contribute to an increased risk of falls.

Due to twisting and arching movements, sitting (and lying) is often a major problem for people with Huntington's disease, as the person frequently tends to come out of a conventional chair/bed. Different specialist chairs (as well as bumpers/cocoons, specialist beds or floor beds) may be required at different stages of the illness, as may positioning harnesses.

A lack of insight, caused by cognitive decline, combined with impulsivity, contributes to an increased risk of falls. For example, a person with severe balance problems may have repeated instances of trying to get out of their chair without asking for assistance. Sensor mats, which are usually placed on the chair or floor, may not be suitable for a person with chorea as the movement may set them off. Self-press falls pendants can be problematic as a person may not appreciate the need to call for help, so passive infrared sensor systems may be more suitable.

Questions To Consider

Managing the risk of falls

- Has the person had falls? If yes, how often? Did they injure themselves and need to seek medical attention or go to hospital?
- Are they having difficulties with involuntary movements, which is leading to a risk of falls? E.g. when they are walking.
- If the person is at risk of falls, how is this being managed? E.g. a falls risk assessment, input from an occupational therapist and/or physio, regular checks and periods of observation.
- What are the risks based on actual incidents/events experienced, as well as potential risks? How have they been identified/assessed and when was the most recent assessment?

Questions To Consider (cont.)

- Does the person have insight into their mobility difficulties and risks associated with falls?
- What other underlying conditions or other Huntington's disease-related symptoms could be affecting the person's mobility?

Equipment and adaptations

- Is equipment being used or adaptations made to reduce the risk of injury? E.g. a hoist to support mobility and assist with transfers.
- If a hoist is used, how many people are required to support this intervention to maintain safety, due to chorea movements, anxiety distress or cognitive impairment?
- Is specialist seating required (e.g. specialist bed/chair) to keep the person safe?
- Does the person use a wheelchair? If yes, under what circumstances? Is it a specialist wheelchair? (I.e. Tilt in Space with built-in pressure relief).
- Are restraints required for safety? E.g. a 3-point harness or lap belt, bedsides and bumpers, passive infrared (PIR) monitoring systems, crash mats. If yes, is there a Deprivation of Liberty Safeguards (DoLS) order in place to authorise this as the least restrictive intervention to promote and maintain safety?

Other mobility issues

- Are involuntary muscle contractions (dystonia) and unusually slow movements (bradykinesia) making it harder for the person to move?
- Is there a risk to self or others due to chorea? E.g. involuntary spasms, hitting out.
- Does the person have days when their mobility is better/worse? If yes, is this linked to sleep or physical ill health?
- Does the person need help with their mobility or to transfer (i.e. to get from the bed to the chair)? If yes, how many people do they need to support them? Do they accept this support and are they aware that they require assistance?
- If the person is experiencing increased pain sensitivity:
  - How is this managed so that it is not a barrier to completing care tasks/maintaining safety and quality of life?
  - Who is responsible for providing that level of support?
  - How successful is the support and how frequently does it need to be reviewed and repeated?
  - What additional knowledge, skills and experience are required to anticipate and interpret needs if the person is non-verbal/ unreliable with communicating needs or symptoms?

6. Communication

As a person with Huntington's disease cannot control the muscles used for speech as well as they previously could, their speech may become slurred, or they may struggle with volume control. This is exacerbated by difficulties in coordinating breathing and speech. They may appear to be shouting angrily, when in fact they are having problems with controlling the volume of their speech.

Cognitive changes in the brain can mean that someone with Huntington's disease struggles to articulate what they are trying to say. This starts early in the disease with the expression/comprehension of complex language. They may find it hard to start a conversation. They may become focused on one topic and not be able to move on from it. This is particularly common if there is a change to the normal daily routine e.g. if there is a clinic appointment and they are worried about getting there. They may also repeat certain words that seem important to them. Ultimately, they may become non-verbal.

A person's communication skills can suffer if they are apathetic, anxious, or depressed. They may not have the drive to organise their thoughts into words, or they may find it hard to be in a group of people. This can mean they start to limit interaction with loved ones, avoid social situations, start to withdraw and feel socially isolated. This can create a cycle of decreasing communication.

Questions To Consider

- Has the person been assessed by a speech and language therapist, who has provided support with communication? If yes, has this been helpful? Was the person able to engage with the advice and intervention?
- Does the person struggle to communicate and tell someone what they need? Would they ask for help if they were in pain or needed something?
- Does the person struggle to understand information and make sense of it? E.g. following instructions.
- How has Huntington's disease affected the person's speech and/or voice? E.g. volume, articulation, word retrieval, clarity.
- What action is being taken to anticipate the person's future communication needs and plan for them? N.B. This is important, as the person will be unable to reliably communicate in the later stages of the disease.

Questions To Consider (cont.)

- Are there other underlying conditions that may also affect communication? E.g. visual deficits, hearing deficits, learning disabilities or having English as a foreign language.
- Is the person prescribed any aids to enable/promote communication, including spectacles, hearing aids, communication aids/devices etc.? If yes, are they able to tolerate using them?

7. Psychological and Emotional Needs

Mental illness is common in Huntington's disease. The most common mental health conditions in Huntington's are depression and anxiety. The person may also experience psychosis.

People with Huntington's disease have a higher risk of suicide than the general population. People with the disease also experience less well-defined, nonspecific changes in personality and mood very early in the illness. This includes irritability, aggression, apathy, or disinhibition, as well as difficulties in understanding social communication.

The central nervous system injury caused by Huntington's disease contributes to these problems. In the early stages of the illness, people may be able to engage with psychological therapies. However, as the disease progresses the complex cognitive requirements of therapy prevent the person from taking part. Pharmacological and less cognitive approaches are often needed if the person will accept them. There is often an interaction between cognitive changes and mood and behaviour i.e. irritability may be caused by an alteration in routine and the cognitive inability to be able to adapt to that change. Social cognitive changes may mean the person is unable to read others' emotions and moderate their own response. Separation anxiety can be expressed through a physical and behavioural response as the person is unable to communicate it in any other way.

Due to the genetic nature of Huntington's disease, people with the condition may have developed unusual ways of relating to others because of growing up in a Huntington's disease family. Many will have had traumatic experiences and developed a trauma response.

Questions To Consider

Psychological needs and mental health symptoms

- Is the person experiencing anxiety, distress, low mood, or perseveration (repetitive ideas)? If yes, has this been assessed? If yes, who carried out the assessment and when did it take place?
- Is the person experiencing suicidal thoughts? Is there a history of suicide attempts? What strategies are in place to reduce the risks associated with potential actions on suicidal ideation or other disordered thoughts?
- Does the person require specific people/routines/items and/or constant reassurance?
- Does the person feel over-stimulated and struggle to cope in busy environments? E.g. shared areas of a care home.
- Are they experiencing neurological apathy? E.g. struggling to get out of bed or initiate tasks.
- Are they socially withdrawing? E.g. avoiding people and activities they would usually enjoy.
- Are they physically withdrawing? E.g. putting themselves on the floor.
- If not provided with encouragement and structure, what would the person do? Is the person highly dependent on familiarity or rigidity in routines, habits, people and environments to feel safe and function?
- Is the person hitting out at themselves or others?
- Does the person have any additional psychiatric diagnoses?
- What happens when it is not possible to have predictability and routine?

Treatment

- Are they adhering to and/or able to cooperate with the treatment/interventions recommended for them? How often is this re-assessed/monitored and who is responsible for ensuring that this happens?
- What are the escalation plans and triggers for any unexpected changes in presentation related to mood/emotional and psychological wellbeing?
- Has the person been admitted to an inpatient mental health service voluntarily and/or under the Mental Health Act?
- Have they received or tried to access psychological support? E.g. therapy and/or counselling.
- Is the person prescribed psychiatric medications as needed (PRN) and/or on a regular basis?
- Is the person under the care of a Community Mental Health Team (CMHT)? If yes, how often are they reviewed by the CMHT and has there been any change in their presentation in the last 3-6 months? If yes, what have these changes been?

Family and relationships

- Does the person become distressed during or after seeing family members?
- Does the person become distressed when seeing people with advanced Huntington's disease?
- Does the person experience guilt because there are children in the family who are at risk of inheriting Huntington's disease?
- Have they observed a parent with Huntington's disease and fear they will end up the same way?

8. Cognition

Due to deterioration in the brain, people with Huntington's disease often find self-monitoring difficult and can lack insight into how the disease is affecting them physically, emotionally and cognitively.

Cognitive difficulties are distinctly different from the pattern seen in other forms of dementia. It is characterised initially by a loss of speed, flexibility in thinking, processing information and poor attention. 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. It also affects executive function and social cognitive functioning. Performance is affected in many areas, including speed, reasoning, planning, judgment, decision-making, emotional engagement, impulse control, temper control, perception, awareness, attention, language, learning, memory and timing. Cognitive impairment will interact with neuropsychiatric, psychological and neurobehavioural challenges.

Detailed cognitive assessment is often only undertaken for Huntington's research purposes or when the clinician is unsure about the presentation. Brief screening tools (such as the Montreal Cognitive Assessment (MoCA)) can be used, although cognitive assessment is often through clinical observation of day-to-day functioning.

Questions To Consider

- Does the person recognise they have Huntington's disease and that they have changed because of it? Or do they not have an awareness of this? Do they have insight into the consequences of the disease and their actions? Can they engage with advice and treatment?
- Can they consent to the assessment process? Can they retain, recall and weigh up the information about the assessment and communicate that?
- Is the person engaging with care to meet their needs or are they experiencing apathy/inertia? Do they need prompting/direction?
- Does the person have poor executive functioning? E.g. do they struggle to plan and multi-task? If yes, how are the risks associated with this managed?
- Even if the person is orientated to people and place and has a good sense of routine, is their decision-making affected? If yes, how are the risks associated with this managed? E.g. prompting, supervision.
- How is the person's memory and ability to process information? If it is poor, how are the risks associated with this managed?

Questions To Consider (cont.)

- Is the person experiencing impulsivity and engaging in risky behaviour? E.g. taking illegal drugs. If yes, how are the risks associated with this managed?
- Has the person been assessed as lacking capacity for any specific decisions? If yes, how has this been managed? (I.e. through a Best Interest process.) Do they have a Lasting Power of Attorney (LPA) for Health & Welfare, who should be consulted?
- Is the person subject to a Deprivation of Liberty Safeguards (DoLS) order? If yes, how is the order being implemented? E.g. actions being taken to stop a person leaving a care home.

9. Behaviour

Behavioural changes in Huntington's disease are common. These changes can be caused by physical changes within the brain. Nonetheless, factors such as reduced independence, strained social life, poor mobility, lack of understanding from others, and the effect that the disease has on employment can cause stress and act as triggers for altered behaviour that challenges.

Behaviours that challenge in Huntington's disease can be immediate and unpredictable, but they can also build up over some time. This is particularly the case with those who cannot or will not engage with any support. This might come in the form of being found in a state of severe neglect or making themselves vulnerable to abuse and not seeing this as problematic.

Questions To Consider

- Does the person respond differently towards different staff and/or family members?
- What triggers behaviour that challenges in the person?
- Does a lack of familiarity with a person's routines and preferences trigger behaviours that challenge towards staff and/or family members?
- Is the person accepting of people visiting them at home?
- Does the person behave in a way that requires intervention to de-escalate? N.B. If yes, record duration, de-escalation techniques used and number of staff involved.

Questions To Consider (cont.)

- Consider the well-managed needs principle: Is the person's behaviour that challenges less of a concern because it is effectively managed by carers and/or family members anticipating their needs and having familiarity with their routine? If yes, how is this behaviour managed? If that management was withdrawn, what might happen? Are there any examples?
- Is the person experiencing side effects from prescribed medication(s), which is affecting their behaviour?
- Does it take time if the person's behaviour changes to identify the reason they are distressed, due to problems with communication? If yes, how is this managed?
- Does the person make involuntary noises?
- Does the person engage in socially and sexually inappropriate behaviour? E.g. disinhibited comments. How is this managed?
- Does the person act impulsively without thinking and not wait for support or assistance? How is this managed?
- Does the person experience neurological apathy? E.g. being withdrawn and distant, lacking interest in things they used to like, and/or not initiating activities, including eating/drinking and hygiene.
- Does the person have a DoLS order? If yes, what has been authorised to be used as the least restrictive options for maintaining safety and wellbeing?
- What knowledge, skills, experience and additional training do professional and unpaid carers need to manage safety in relation to the person's risk-taking behaviour?
- How often are carers and/or family members still 'on the back foot' and needing to interpret and respond to sudden/unexpected changes in presentation and behaviour, in spite of current care plans and risk assessments? How do they 'manage' unpredictable behaviour?
- Are medications or physical restraint ever needed as an immediate response to prevent serious harm to self or others?
- Is the person doing something that increases their risk of harm from other people who may be distressed by their behaviour?

10. Drug Therapies and Medication: Symptom and Control

There is currently no 'cure' for Huntington's disease. Medications are prescribed to manage symptoms, but these are not always effective or cause other side effects. For example, a drug prescribed to minimise movement, may increase problems with cognition and dampen mood, particularly in those prone to depression and suicidal ideation.

Whilst people are often hyposensitive to pain, there is a type of pain known as Huntington's disease pain (in the hips/rear of thighs) which can prove particularly difficult to treat. To manage pain towards the end of life, people with Huntington's disease may require additional or increased doses of medication, which can fall outside of the usual licensing of the chosen drug.

Treatments for issues such as contractures (i.e. baclofen) may not be effective. Botox for spasticity may not be possible to administer due to the intensity of movement. Hypersensitivity to anti-secretion medications is common.

As the disease alters, so do the symptoms. Knowing the person's profile and history is the key to assessing change and triggers, particularly when cognition and communication is so impaired.

Huntington's disease teams acknowledge that the disease is palliative and will make attempts to ensure advance planning is in place. However, this is not always possible due to denial, cognitive impairment, or an unwillingness to discuss it. Palliative care and treatment may frequently have to be discussed under the terms of the Mental Capacity Act.

Questions To Consider

- What type of medication is the person prescribed? N.B. It is common for people with Huntington's disease to be prescribed anti-psychotic medication, which GPs can be reluctant to prescribe.
- If the person is prescribed more than one medication, how frequently are medications being monitored and evaluated for efficacy and side effects?
- What form of treatment does the person's medication take? E.g. tablet, oral liquid, PEG.
- What support does the person require to take their medication? Is the person compliant with their medication? Does administration of medication require multiple attempts to gain co-operation? If yes, record number of attempts and time taken on average. Is there a need for 'covert' medication and an associated capacity assessment and Best Interest Decision?
- Has the person been able to engage in any form of advance planning?

11. Altered States of Consciousness (ASC)

People with Huntington's disease are at increased risk of brain injury, often due to unwitnessed falls. This can lead to altered states of consciousness from their baseline presentation i.e. hypersomnia. It is not uncommon for children/young people with Huntington's disease to experience seizures as well as people with (adult) advanced Huntington's disease. These should be managed in line with standard epilepsy treatment protocols.

Questions To Consider

- Does the person experience seizures? If yes, how are these seizures being managed?
- What other conditions or symptoms may also influence consciousness levels? E.g. risk of ASC associated with falls and head injury, or stroke/ TIA (transient ischaemic attack) risk, in addition to seizures.
- Are there any prescribed medications, which may have side effects, which affect levels of consciousness?
- Are abnormally high/low blood sugar levels likely to be a factor in symptom management?

12. Other significant care needs

Huntington's disease affects a person's ability to carry out their activities of daily living. Subtle personality and cognitive changes often begin many years before movement symptoms. There are many different presentations of the illness. CHC Assessors should be aware that the main changes are executive dysfunction, social cognition and neuro-behavioural change (particularly apathy). These changes may lead people to struggle with decision-making, relationships, parenting, managing a home, managing their finances and maintaining employment.

Partners, companions and children of people living with Huntington's disease often take on the burden of these invisible changes. They may not wish to draw attention to the changes, being concerned about the person's response. As the person with Huntington's disease may be struggling with insight into these changes, CHC Assessors are advised to listen to the companion's history of any changes and report of their day-to-day function and interaction. This conversation needs to be initiated sensitively and may need to take place away from the person living with Huntington's disease.

Questions to consider are:

- Is the person struggling with day-to-day activities and interaction?
- What is the timeline and history of the changes for the person with Huntington's disease and their companion or supporter? Is the history given by the two parties different?
- What social and non-social functions has the companion taken on from the person with Huntington's disease?

Once considered solely a movement disorder, Huntington's disease is now considered a neuropsychiatric, degenerative illness with a range of presentations (known as 'phenotypes'). It is also now recognised as a systemic disease with many of the symptoms interacting with one another.

Questions to consider are:

- What is the interaction of different areas of need? For example:
  - How do a person's communication difficulties affect their behaviour if they are unable to communicate the cause of their distress?
  - Can a person tolerate an increase in prescribed supplement to address the healing of a wound without vomiting and causing them to withdraw from treatment?
  - Will their cognitive impairment allow them to comprehend how to use a mobility aid to reduce the risk of falls?
  - What level of knowledge and skill is required to manage and support the person with Huntington's disease?
  - What amount of resources and time are needed to address the person's needs? Does the person understand the level of support needed?

Get in touch

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

email **info@hda.org.uk**
phone **0151 331 5444**

www.hda.org.uk

Get involved

For your fundraising pack, please get in touch with the fundraising team

email **fundraising@hda.org.uk**
phone **0151 331 5445**

Join the conversation

Huntington's Disease Association
Liverpool Science Park IC1,
131 Mount Pleasant, Liverpool L3 5TF

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