Huntington's Disease Association

Speech and language therapy for people living with Huntington's disease

RCSLT

EHDN EUROPEAN HUNTINGTON'S DISEASE NETWORK

Advancing Research, Conducting Trials, Improving Care

Registered Charity No. 296453

Contents:

Authors

Erin Probert is a Clinical Lead Speech and Language Therapist at the Royal Hospital for Neuro-disability, London. She has extensive experience working with people with Huntington's disease in a specialist inpatient setting, supporting them with communication, swallowing and decision making.

Sarah Roberts is a Highly Specialist Speech and Language Therapist working in the NHS, with a special interest in progressive neurological conditions. Sarah has been working with adults with neurological conditions for the past 10 years and has worked across both inpatient and community settings.

Community reviewer

Allan Adams

Introduction

This guideline covers the role of speech and language therapy in the management of Huntington's disease. It is recommended that speech and language therapists (SLT) working with someone with Huntington's disease use the resources on the Huntington's Disease Association's website to familiarise themselves with the general features of the disease before reading this guideline.

The guideline has been developed by reviewing the available evidence and expert consensus on best clinical practice. It is broadly based on the special reports produced by the European Huntington's Disease Network Standards of Care Speech and Language Therapy Working Group on oral feeding and communication in 2012 (Hamilton et al). Although these papers are now over a decade old, they remain the most recent expert consensus summary of the SLT role in supporting people with Huntington's disease. This guideline is intended to provide a starting point from which SLTs can complete further reading as required.

People with Huntington's disease will develop dysarthria (challenges speaking clearly because of difficulties with the muscle movements needed for speech), cognitive-communication impairments, and challenges with social interactions. This broadly aligns with the 'triad of impairments' within Huntington's disease, where the person experiences motor disorder, cognitive disorder and develops neuropsychiatric features. The combination of these changes has a profound effect on people's ability to express

themselves and maintain relationships. People with Huntington's disease will also experience a deterioration in their swallowing function. This will lead to challenges with maintaining adequate nutrition and hydration, risks associated with aspiration and choking, and a negative impact on their wellbeing.

Skilled assessment and management of communication and swallowing are essential at each stage of the disease. This is because of the need to maximise the person's abilities, maintain independence and autonomy, promote the best possible quality of life, and help manage risks. It also helps people to retain function for as long as possible.

At every stage, it is vital that SLTs provide patient-centred, holistic care. This means there should not be a sole focus on physical symptoms. Consideration should also be given to emotional, social, psychological, spiritual and cultural needs and preferences. This approach emphasises that the person is an active participant in their own care and allows them to make their own informed decisions about treatment and therapy.

Part 1 - Communication

How Huntington's disease affects communication

The following section sets out the core ways in which Huntington's disease affects communication. However, each person will have specific communication strengths and needs. A careful, individualised approach to assessment and management is needed.

Dysarthria

The choreic movement disorder that most people with Huntington's disease exhibit affects the muscles needed for speech production. Huntington's disease particularly impacts the basal ganglia, and therefore, the pattern of dysarthria is generally hyperkinetic. However, some people have a more rigid muscular presentation, where paucity of

movement and stiffness are common.

The common features of dysarthria in Huntington's disease are outlined below (Diehl et al, 2019; Hamilton et al, 2012):

- Irregular breakdown of articulation, affecting consonants and vowels
- Lack of rhythm with pauses within and between words, prolonged phonemes (lengthening of speech sounds)
- Mistiming in breath control with erratic and variable inspiration and expiration
- Reduced coordination of respiration, phonation, and articulation
- Inappropriate silences
- 'Strained - strangled' voice quality
- Variability of volume control, may present with monoloudness
- Reduced or inappropriate prosody (e.g. stress / intonation)
- Changes to speech rate, may present as abnormally fast or abnormally slow

In early-stage Huntington's disease, the person is likely to have mild dysarthria only, which may impact the precision of their articulation and rhythm of speech.

Speech is likely to still be fully intelligible. As the disease progresses, their dysarthria will worsen, with speech intelligibility deteriorating until, in the later stages, the person is not able to communicate effectively via speech at all.

People with earlier-stage Huntington's disease have described some of the challenges related to their speech, such as mistakenly being thought to be drunk, or believed to be angry because they are 'shouting'.

Cognitive-linguistic impairment

The steady decline of cognitive function in people with Huntington's disease will impact language and broader cognitive-communication skills. There is a complex interplay between language impairment and cognitive impairment, meaning it may not be possible to differentiate between them. Hamilton et al (2012) identify many of the key features, which are outlined below:

- Increasing difficulty with language comprehension – this starts as problems with 'higher' level functions, such as understanding inferences, and gradually worsens

- Diminishing use of broad and varied vocabulary, with a reduction in the number / range of words used
- Increasing difficulties with word finding
- Reduction in sentence / phrase length
- Reduction in complexity of sentence structure
- Difficulty generating ideas and expressing them in an orderly and coherent way
- Difficulty with topic maintenance
- Perseveration on words, phrases, topics or thoughts
- Increase in processing time

Challenges with social interaction

The cognitive and neuro-psychiatric elements (such as depression and anxiety) of Huntington's disease interplay to cause difficulties with social interaction. The person with Huntington's disease may experience:

Changes in social behaviour, such as:

- **Apathy:** This can present as a reduction in emotional responsiveness, low levels of motivation, and withdrawal from interacting with others
- **Challenges with initiation:** This includes finding it difficult to start or continue a conversation
- **Frustration and irritability:** This is linked with difficulties regulating emotions
- **Impulsivity:** This can present in conversation as not allowing the other person to speak or abruptly changing topic
- **Behaviours that challenge:** An example is physical or verbal aggression
- **Difficulties with adapting speech content to the situation:** This can lead to saying things that could be perceived as inappropriate
- **Difficulties in maintaining a socially acceptable level of proximity:** This can lead to them getting very close to the person they are interacting with
- **Difficulties in maintaining attention on the other person**

Changes in non-verbal communication, such as:

- Choreic movements impacting body language, facial expressions and maintenance of eye contact. This can affect how the person is perceived and lead to misinterpretations of their mood or intent

- Challenges using gestures to support communication due to a lack of coordination and control of movements
- Difficulty using Alternative and Augmentative Communication (AAC)

People may also present with reduced insight into their communication difficulties, which can have a significant impact on their motivation and ability to use communication strategies or aids.

Due to these challenges, the person may start to struggle in social situations, particularly those that are busy and involve lots of people. These symptoms can be experienced even at an early stage of Huntington's disease. Over time, this can lead to the person avoiding contact with others. This risks increasing social isolation, leading to a reduction in communication opportunities (such as meeting friends), which are necessary to maintain social skills.

Speech and language therapy management approaches

The approach needed to manage these difficulties varies hugely over the course of the disease and is dependent on the needs, wants, and priorities of the person. The focus and goals of intervention should be discussed and agreed with the person themselves, or if they are not able to take part in this, through liaison with family members where possible.

There is a lack of high-quality evidence for many of the interventions outlined below. The information shared is not intended to be exhaustive or prescriptive.

Multi-disciplinary team working

It is recommended that interventions for people with Huntington's disease are provided as part of a multi-disciplinary team (MDT) approach, as this is more likely to meet the needs of the person in an effective way. The interplay between the physical, cognitive, and neuropsychiatric elements of Huntington's disease means that collaborating with MDT colleagues, where possible, makes interventions more likely to be successful. SLTs can also support other members of the MDT with how to best optimise the person's understanding and expression during their interactions. MDT members may include:

- **Physiotherapy:** can support with postural management to facilitate communication, optimisation of breath support for speech and positioning for AAC access
- **Clinical psychology:** can help with optimising cognitive and behavioural aspects of Huntington's disease, to support interactions
- **Occupational therapy:** can aid access to enjoyable, motivating and social activities, and support management of cognitive changes
- **Medical team:** for pharmacological management of symptoms that are impacting communication and advice on possible medication side effects of relevance (such as antipsychotic medication potentially worsening the presentation of dysarthria – Rusz et al, 2014)
- **Carers:** integral in providing opportunities for social engagement and supporting the person's communication success

Early-stage management

The person may present with mild dysarthria and mild cognitive-communication impairments at this stage. They may be experiencing some changes in their interaction style and subtle changes in behaviour, which might be starting to affect their relationships. Challenges in regulating emotion can have a significant impact on effective communication (Zarotti et al, 2017).

Things to consider at this stage include:

- **Early referral for speech and language therapy:** This will allow initial assessments of communication, exchanging of information and advice, and development of therapeutic relationships between the speech and language therapy team and the person and their family. Grimstvedt et al (2021) highlight that in view of the 'dramatic impact of Huntington's disease on patients' communication skills', SLTs should ideally be involved from the early stages of the disease, while the patient is still capable of voicing his / her own wishes and thoughts'. Early referral to an SLT is also recommended by Bachoud-Lévi et al (2019) in their International Guidelines for the Treatment of Huntington's Disease, with reference to the role of SLTs in supporting communication throughout the disease, and the

changing needs of the person

- **Informal and formal assessments:** Assessment of the person's speech can be done both informally (e.g. in conversation) and by using a formal dysarthria assessment tool (e.g. the Frenchay Dysarthria Assessment). Similarly, assessment of the person's receptive and expressive language abilities can be completed both informally and using more formal assessments (such as the Mount Wilga High Level Language Test or the MCLA - Measure of Cognitive Linguistic Abilities). This can help to identify any breakdown in comprehension of complex or abstract information, or other 'higher level' language functions that involve more complex processing (Saldert et al, 2010). Assessment at this point both provides information to guide early input in maximising strengths and compensating for weaknesses, and also to establish a baseline against which to measure change
- **The impact of any communication difficulties:** Spending time identifying the person's specific priorities and concerns is important. People with Huntington's disease and their loved ones may perceive the main challenges around communication differently (Hartelius et al, 2010). Therefore, it is useful to involve the people around them (where the person gives consent to this) in building up a picture of their communication needs
- **Intervention approach:** Early on, some people might benefit from a rehabilitation approach (Zinzi et al, 2007). For dysarthria, this could include laryngeal relaxation techniques for voice, exercises to work on coordination of respiration and phonation, intonation drills, and work on pacing of speech. Another approach is compensatory strategies like optimising the environment and using conversational repair strategies, such as rephrasing the message. Consider how generalisation of any intervention or strategies into functional and meaningful activities will be achieved. The person may also benefit from work on improving self-monitoring of speech to allow them to more easily compensate for any changes affecting intelligibility
- **Information and advice provision:** This is likely to be helpful for the person with Huntington's disease and their conversation partners to help facilitate understanding of communication challenges and strengths, how to support their understanding and self-expression, and maintenance of social interactions
- **Future wishes:** It is important for decision-making about future wishes regarding care and treatment (such as options for nutrition / hydration, where they would like to live, etc.) to be raised by the MDT, while the person has the cognitive and communicative capacity to engage in this.

The person may express that they are not ready to talk about it or do not want to. However, it is vital that it is offered, and the person knows that they can make their wishes known

- **Voice banking:** Some people with early (or even pre-symptomatic) Huntington's disease may wish to consider voice banking. This is not something very commonly used for people with Huntington's. Be aware that the person may not be able to access voice output high-tech Alternative and Augmentative Communication (AAC) at the point that they would need to use it. However, voice banking can be an empowering step that people can take in preserving their identity. Message banking and legacy recordings can also be considered and may be more appropriate

Mid-stage management

As the disease progresses, research shows increasing challenges in communicating effectively and being able to actively listen to others (Watson et al, 2021). Speech intelligibility will continue to deteriorate – usually dysarthria worsens in line with the progression of the disease (Hartelius et al, 2003). Cognitive changes mean the person will find new learning increasingly difficult. They will likely find it harder to learn, recall and initiate any strategies to support their interactions. Their insight into their communication challenges is also likely to decline. This increasingly means that communication partners need to be the ones to use strategies and supports to help facilitate communication and maximise the success of interactions.

Things to consider at this stage include:

- **Conversation partner training:** This can be a very helpful approach when tailored to the person with Huntington's disease. Environmental factors, such as family support, can be both key barriers and facilitators to communicative participation (Power et al, 2011). People with Huntington's disease have described how the communication style of others can impact on communication negatively, such as when people are speaking too quickly (Hartelius et al, 2010)
- **Re-assessment:** Consider whether this is likely to yield any additional information that may change management or advice, beyond that gained from the informal assessment that happens naturalistically in interactions

and from patient / family feedback

- **Talking Mats:** As the person's ability to make themselves understood verbally declines, and their cognitive impairments increase, a useful tool to help with self-expression and decision-making is Talking Mats. This is one of the few approaches for the speech and language therapy management of Huntington's disease that has a more significant evidence base (e.g. Ferm et al, 2012, 2010; Hallberg, 2013)
- **Total communication approach:** This encompasses both the person and their communication partners using any and all forms of communication to support comprehension and expression, e.g. gestures, drawing, objects, writing. Even when the person with Huntington's disease finds it difficult to use some of these strategies themselves (e.g. if legible writing is not possible due to choreic movements), it is likely that they will still be helpful for others to use with them, to support their processing and understanding
- **Alternative and Augmentative Communication (AAC) options:** Many people with Huntington's disease may have the ability to use AAC methods. However, the use of AAC is not consistently explored or trialled for people with Huntington's disease (Grimstvedt et al, 2021). AAC can encompass 'low tech' AAC (such as picture charts, symbol-based communication books), 'light tech' AAC (such as switches that play recorded messages, simple text-to-speech devices such as lightwriters) and 'high tech' AAC (such as text- or symbol-to-speech apps on a tablet). The AAC that the person can access will change over time and will be impacted by their complex and sometimes variable presentation of symptoms (Diehl et al, 2019). Some people may present earlier with more severe dysarthria whilst maintaining relatively intact cognitive function. If so, they may be able to use more complex AAC options to support themselves. Others may require very simple, low-tech charts. The AAC offered must be appropriate for their abilities and stage of the disease. Consider how the person with Huntington's disease will physically use any AAC, in view of the uncontrolled movements they are likely to be experiencing. Some people with Huntington's disease will meet the criteria for support from an NHS AAC hub, which can also support with access requirements
- **AAC timing:** General guidance around the introduction of AAC devices is to introduce them early, when the person is still able to learn how to use them. This is in the hope that they will then be better able to utilise it as the disease progresses. Whilst this may be sound in principle, it can be difficult in practice to engage and motivate the person in using AAC

before they absolutely need it

- **Ongoing opportunities for interaction and social engagement:** This should be supported as much as possible – for example, by advising on adapting usual social events to include smaller groups of people or to take place in a different environment. It can also be beneficial to explore local or online peer groups. There is evidence that people with Huntington's disease find it easier to communicate in situations where they feel safe and comfortable (Zarotti, 2017)
- **Capacity assessment:** SLTs have a key role in supporting the assessment of mental capacity. In order to comply with the Mental Capacity Act (2005), 'all practicable steps' need to be taken to help someone to make their own decision before it can be deemed that they lack capacity. For someone with a complex communication impairment, such as those arising from Huntington's disease, speech and language therapy support can be vital. SLTs can use their expertise in facilitating communication to help the person with Huntington's disease to understand, weigh up and communicate their decision. Even when a person is found to lack capacity for a certain decision, SLTs can play an important role in advocacy for the person during the best interests process. For example, by sharing what information or views the person is able to express. Talking Mats, as referred to above, can be very helpful as part of a capacity assessment for someone with Huntington's disease

Late-stage management

In the late stages, the person with Huntington's disease is not likely to be able to communicate intelligibly using speech, and their cognition will be severely impaired. They may have some basic comprehension of simple information, particularly when supported with visual cues and context. Their ability to use AAC or other compensatory techniques, such as gestures or writing, is likely to be severely diminished or impossible. The person may also have significant difficulties with initiation and have low frustration tolerance.

Things to consider at this stage include:

- **Utilising residual skills:** It may still be possible for the person to use a simple communication system – such as thumbs up / down or head nod / shake for yes / no responses, or to make simple choices from two items
- **Communication opportunities:** Maximising opportunities for positive social interactions and experiences, and supporting and training communication partners
- **Guidelines:** Advice to support and facilitate communication should be clearly documented and conveyed. The aim is for the person to maintain some autonomy for as long as possible

Part 2 - Swallowing

How Huntington's disease affects swallowing

Huntington's will affect a person's eating, drinking, and swallowing as the disease progresses. Dysphagia is prevalent in 35% of early-stage, 94% of moderate-stage and 100% of advanced-stage Huntington's disease patients (Schindler et al, 2020). However, information on best management is significantly limited (Pizzorni et al, 2020). Aspiration pneumonia is the leading cause of death for people with Huntington's disease (Pizzorni et al, 2020). Therefore, SLT input is integral as part of the overall management of dysphagia within the Huntington's disease population.

The information below has been written as a brief guide for SLTs and is largely based on the special reports from the European Huntington's Disease Network Standards of Care Speech and Language Therapy Working Group (Hamilton et al, 2012) and a more recent narrative review by Sapmaz et al (2024).

Due to the complex interplay of physical, cognitive, and behavioural symptoms of Huntington's disease, it is essential that the approach taken to manage dysphagia is holistic in nature. In practice, this means looking at the whole person in their own specific situation and environment. SLTs have a key role in supporting the person and their carers in managing risks associated with dysphagia, whilst optimising and promoting quality of life.

Physiological changes and impact on swallowing

Huntington's disease affects the brain areas responsible for motor control, leading to involuntary movements (chorea) that can disrupt the coordinated movements needed for swallowing. Other motor impairments include muscle stiffness (rigidity), dystonia (involuntary and prolonged muscle contractions) and slowness of movement (bradykinesia). These motor impairments affect the person's swallowing function.

Hamilton et al (2012) describe some of the key swallow features in Huntington's disease:

- Hyperextension of neck and trunk
- Reduced mastication and lingual control
- Darting lingual chorea
- Drooling – spillage from mouth
- Very rapid eating
- Premature transfer, particularly of liquids
- Intraoral retention following initial transfer
- Delayed and repetitive swallow
- Prolonged laryngeal elevation
- Coughing and choking
- Reduced / disrupted breath control during the swallow
- Phonation during swallowing
- Belching
- Aerophagia
- Vomiting

Choreic movements can affect the person's limb control, meaning they find it difficult to get food or drink to their mouth and may have difficulty taking reasonable-sized mouthfuls.

Medical treatment for chorea can sometimes help to improve swallowing problems. However, consider the potential side effects of treatments for chorea (e.g., sedation, attention, and Parkinsonism), which might also impact

on the person's swallowing ability (Bachoud-Lévi et al, 2019). Some medications may also induce dry mouth (Rada, 2008).

Cognitive changes and impact on swallowing

The impact of Huntington's disease on cognition and behaviour can also have a significant impact on swallowing, including:

- **Difficulty engaging with recommendations:** People may find it difficult to understand the rationale for speech and language therapy recommendations, or to engage in discussion about management options. This can mean they are reluctant to follow risk-reduction measures
- **Difficulty initiating tasks:** The person may have difficulty initiating tasks, such as eating and drinking, and swallowing. This can often be supported by cueing or prompting the person to initiate the task
- **Fixation on objects / events:** Cognitive and behavioural changes in people with Huntington's disease may lead to a refusal of particular foods or fixation on certain foods. The person may also become hyper-focused on a range of different things. This can include fixation on specific utensils, leading to not being able to accept different ones, fixation on eating a certain food repeatedly, or fixation on certain ways of living, including eating healthily
- **Problems maintaining attention:** Difficulties with sustaining or switching attention can lead to the person losing focus on their meal, causing them to become distracted. This can affect how much they eat / drink and increase the risks of aspiration and choking
- **Impulsivity:** People may have increased difficulty with impulsivity. This can result in difficulty controlling the amount of food they put into their mouth at one time, which can lead to overfilling. Impulsivity can also lead to an increase in the speed of eating. Impulsivity can be a significant factor in increasing the risk of choking, aspiration, oral residue and anterior loss of food from the mouth
- **Behavioural changes:** For some people with Huntington's disease, stressors around mealtimes can be a particular trigger for behaviours that challenge. For example, they may become frustrated and refuse to eat, or may throw food or cutlery

Other considerations

SLTs should consider the following when assessing and managing symptoms in a person with Huntington's disease:

- **Mood and wellbeing:** SLTs should be mindful of how mood can affect appetite, such as if the person is experiencing anxiety or depression. Conversely, they should also be aware of the potential for problems with swallowing to influence someone's wellbeing. For example, the embarrassment that someone may feel if they 'make a mess' when eating and drinking, or the person having difficulty maintaining the social aspects of eating and drinking. Sapmaz et al (2024) report that dysphagia in Huntington's disease can lead to social isolation and a diminished quality of life
- **Weight loss:** Swallowing difficulties can lead to weight loss as eating becomes more challenging and less enjoyable. The person may struggle to meet their significant energy requirements, which are exacerbated by choreic movements. Therefore, close working with the dietetics team is vital
- **Saliva management:** Saliva management can be problematic for people with Huntington's disease, often due to a build-up of saliva caused by impaired swallowing and poor oral control (Bachoud-Lévi et al, 2019). SLTs should be mindful of the possible side effects from anticholinergic medications used to reduce saliva, being aware that these medicines may negatively impact cognitive function and alertness. The use of salivary gland botulinum toxin injections can be considered. However, their use may not be possible due to the risk associated with uncontrolled body and head movements. SLTs should liaise with the medical team about saliva management options

Speech and language therapy management approaches

The information below gives some general principles. Advice and management should always be tailored to the person and their needs.

Although there are some commonalities in the presentation of Huntington's disease, its specific impact varies from person to person and changes over time. This is why an individualised approach is always needed.

Assessment

Early referral to an SLT is recommended to allow timely assessment and advice giving. SLTs can also help to address specific anxieties and concerns that may arise if the person with Huntington's disease has experience of family members with the same condition.

Conduct a thorough assessment, considering not only the person's dysphagia presentation but also wider environmental factors. To this end, assessment during a mealtime in the person's usual environment can be very helpful, where possible. This is because only seeing the person in an isolated context, such as in a clinic, can mask some of the difficulties that they might be experiencing. Detailed interview with the person and, with their consent where possible, their family members or carers, is essential.

Pizzorni et al (2020) discuss the use of instrumental assessment of swallowing, such as videofluoroscopy or fibreoptic endoscopic evaluation of swallowing (FEES), to aid diagnosis of dysphagia in Huntington's disease patients. However, this may be more difficult if the person cannot participate due to involuntary movements or cognitive issues (Hamilton et al, 2012). Therefore, it is often preferable to carry out this assessment at an early stage. Be aware that instrumental evaluation can only provide information on a snapshot in time and that swallowing will change as the disease progresses.

Multi-disciplinary team working

Multi-disciplinary team (MDT) working ensures holistic and collaborative care, which is a necessity when attempting to reduce dysphagia symptoms (Sapmaz et al, 2024). The MDT can also provide vital support to the person and to their family when considering advance care planning and decision-making.

Other members of the team may include:

- **Occupational therapy:** This is useful when seeking advice on specialist equipment / utensils, and on maximising independence with eating and drinking

- **Physiotherapy:** This can help with advice on specialist seating and positioning to help with safer swallowing
- **Dietetics:** This can help with the maintenance of adequate nutrition / hydration and a healthy weight
- **Clinical psychology:** This can help with managing the cognitive and behavioural elements of Huntington's disease to maximise mealtime safety and enjoyment
- **Nursing:** Depending on the setting, nursing support may be integral in helping the person day-to-day with oral intake and ensuring any concerns or changes are reported
- **Medical team:** This can be useful for pharmacological management and non-oral feeding

Non-oral feeding

It is vital that non-oral feeding is discussed for people with Huntington's disease. This is due to the deterioration of swallow function as the disease progresses. This leads to the person being at high risk of aspiration and choking on all textures and unable to maintain adequate nutrition and hydration orally. Support the person to consider whether they would like a gastrostomy for the administration of clinically assisted nutrition and hydration (CANH) (with or without some ongoing oral intake for enjoyment). The alternative would be to continue with solely oral intake. Continuing with solely oral intake will inevitably become eating and drinking with acknowledged risks (EDAR), which can be further supported by the SLT and MDT (RCSLT, 2021). Bachoud-Lévi et al (2019) discuss that the aim of non-oral feeding is to improve quality of life rather than increase the length of life.

Ensure that the person has an opportunity to make this decision, whilst they retain the cognitive and communicative ability to express their wishes and preferences. Ideally, discussions regarding advance care planning would take place while the person retains the mental capacity to make their own decision. SLTs have a role in providing information about dysphagia and management options, supporting mental capacity assessments, and participating in best interests decision-making when indicated. Even if the person lacks the mental capacity to make the decision themselves, supporting people with communication challenges to understand the relevant information and make their thoughts known can be invaluable in

bringing the person's own perspective into decision-making.

Some people with Huntington's disease may not want to engage in decision-making at this early stage, which is their right. They may not feel comfortable talking about the disease's progression, which is an inextricable part of planning for the future. However, they should be made aware that they can raise this at any time and provided with information about how to do this.

If the person does make a decision, whether that is to have a gastrostomy placed in the future or not, ensure they are aware that they can revisit this decision at any point should they wish to.

Changes to case law regarding CANH discontinuation and the subsequent publication of British Medical Association guidance (2018) have changed the landscape of decision-making around CANH. The guidance makes it clear that CANH can be discontinued in advanced Huntington's disease. This allows the person to die without the need to go to court, provided everyone is in agreement that this is in the person's best interests. Therefore, if the person makes a decision to have a gastrostomy tube, it is important for their wishes to be documented, regarding whether there is a future point when they may like CANH to be stopped.

The SLT and broader MDT should be aware of relevant legal tools such as Advance Decisions to Refuse Treatment (ADRT) and Lasting Powers of Attorney (LPA) for Health & Welfare. Consider informing the person with Huntington's disease (providing they have mental capacity) about these tools, explaining their potential relevance when someone lacks the capacity to make their own decision about CANH / EDAR.

Early-stage management

Hamilton et al (2012) report that early on, there may be no obvious signs of dysphagia or subtle signs, including occasional coughing when eating / drinking. There may be slight difficulties in mastication and reduction of lingual control.

Management at the very early stage may mostly incorporate education and advice, and can include:

- Advice on managing any difficulties the person is having with foods / fluids – such as avoiding challenging textures or adapting foods, moderating mouthful size, etc

- Advice on managing any anxieties around choking, or embarrassment around drooling or food spillage
- Advice on resting before and after meals to conserve energy
- Advice on environmental adaptations – such as reducing distractions and ensuring that the environment is quiet and relaxed. Advise on relaxation techniques to help relieve tension when eating and drinking

Mid-stage management

Mid-stage Huntington's disease can coincide with the first signs of swallowing difficulty or a significant exacerbation, with dysphagia having a greater impact on the person's ability to maintain a healthy weight, medical stability, and wellbeing (Hamilton et al, 2012). MDT working becomes increasingly important.

The SLT should continue to provide reassessment should there be any changes to the person's swallowing function. SLTs also need to consider the cognitive and communication changes that a person may be experiencing, recognising the impact this can have on the management of their eating and drinking. This includes the ability to understand and follow recommendations, which will decline as the disease progresses.

At this stage, people may have difficulties with self-feeding and monitoring feeding rate (Keage et al, 2020). This can lead to overfilling their mouth, increasing the risk of choking or anterior loss of food.

Develop a management plan in partnership with the person and their caregivers. It should consider:

- Placement of the bolus, its size and pacing of feeding
- Modification of food and fluid textures as per International Dysphagia Diet Standardisation Initiative (IDDSI) descriptors – whilst acknowledging the risks and benefits of each option
- If the person is expressing a desire to eat and drink with acknowledged risks (EDAR), then this should be supported and explored (RCSLT, 2021). A capacity assessment is likely to be required
- Trialling different postures or swallow manoeuvres to optimise swallow safety and reduce choking and aspiration risk. This can include optimising head positioning to support the person remaining in a neutral position

and to combat hyperextension

- Offering meals when the person is most alert
- Considering smaller, more frequent meals to reduce fatigue
- Encouraging self-feeding for as long as possible to maintain independence. Consider the use of specialist cutlery or cups, such as flow rate control beakers
- If the person becomes fatigued and is unable to feed themselves, consider hand-over-hand assistance initially if possible. This is usually preferable to going straight into feeding the person
- Providing education and training to caregivers on how to optimise mealtime enjoyment and safety, with a clear rationale for recommendations to help support compliance
- Providing education to caregivers on how to help the person initiate swallowing, such as the provision of an empty spoon if holding food in the mouth, verbal and visual reminders to chew and swallow food
- Liaising with occupational therapy and physiotherapy to optimise posture and positioning
- Close working with dietitians to ensure patients are meeting nutrition and hydration requirements, particularly if movements are increasing and diet is being modified

Late-stage management

As the disease progresses, maintaining adequate nutrition and hydration following further changes in cognition, mood, communication and appetite becomes increasingly challenging (Hamilton et al, 2012; Sapmaz et al, 2024).

The person will be at an increased risk of aspiration and choking, and may experience associated health complications such as chest infections. Hamilton et al (2012) report that the cough reflex is well preserved. However, Schindler et al (2020) found silent aspiration in 27.8% of advanced-stage Huntington's disease patients, compared to 7.7% and 11.8% at the early and moderate stages, respectively. They also found a strong correlation between disease progression and dysphagia severity, associated with worsening of motor symptoms. In addition to chorea, the presence of rigidity and dystonia is higher in the later stages of Huntington's disease (Sapmaz et al, 2024).

The person is likely to be dependent on others for both feeding and oral hygiene – this increases the risks associated with aspiration (Langmore et al,

1998). Maintaining optimal oral hygiene can become more difficult due to involuntary movements, impaired cognition and behaviours that challenge.

Management at this stage should be adjusted in accordance with the person's cognitive level (Sapmaz et al, 2024) and will vary depending on decision-making about non-oral feeding and EDAR.

Management may include:

- Regular review of swallowing
- Continued review of dietary modifications and compensatory strategies to support safe and efficient swallowing, whilst ensuring that the person's prior expressed wishes and quality of life remain key considerations. This stage may be where EDAR is considered as a management option in conjunction with discussions within the MDT
- Further discussions about non-oral feeding – this may include starting or stopping this
- Optimising oral hygiene as much as possible
- Caregiver training and education about how to support the person

Conclusion

Both communication and swallowing impairments have a major impact on the lives of people with Huntington's disease and their families. SLTs have a vital role in supporting people with Huntington's, using a person-centred and multi-disciplinary approach. This will help the person maintain autonomy for as long as possible and facilitate the best possible quality of life, whilst managing risks.

Acknowledgements

This guideline has been produced by the Huntington's Disease Association and is endorsed by the Royal College of Speech and Language Therapists - the UK's professional body for speech and language therapists.

It is based on the special reports produced by the European Huntington's Disease Network Standards of Care Speech and Language Therapy Working Group on oral feeding and communication in 2012. We would like to thank all of the authors and contributors to the EHDN guidelines.

Resources

Huntington's Disease Association resources for professionals

The Huntington's Disease Association offers a range of resources to support professionals working with people affected by Huntington's disease.

You can find our resources at **www.hda.org.uk**

Explore the professionals webpage using the search bar to find specific resources. Search "resources".

Stay informed

Join our professional-only mailing list to receive updates on events, webinars, and new resources tailored to your needs. You can sign-up at **https://www.hda.org.uk/professionals-and-training/**

Support for children and young people

Find information on our Huntington's Disease Youth Engagement Service (HDYES) by searching for "HDYES" on the website.

References and further reading

Communication

Bachoud-Lévi, A-C., Ferreira, J., Massart, R., Youssov, K., Rosser, A., Busse, M., Craufurd, D., Reilmann, R., De Michele, G., Rae, D., Squitieri, F., Seppi, K., Perrine, C., Scherer-Gagou, C., Audrey, O., Verny, C., Burgunder, J,M. 2019. International Guidelines for the Treatment of Huntington's Disease. Frontiers in Neurology.

Diehl, S., de Riesthal, M. 2019. Augmentative and Alternative Communication Use by Individuals With Huntington's Disease: Benefits and Challenges of Implementation. Perspectives of the ASHA Special Interest Groups.

Department for Constitutional Affairs. 2007. Mental Capacity Act 2005: Code of Practice. London: TSO.

Diehl, S. K., Mefferd, A. S., Lin, Y. C., Sellers, J., McDonell, K. E., de Riesthal, M., Claassen, D. O. 2019. Motor speech patterns in Huntington disease. Neurology.

Ferm, U., Eckerholm Wallfur, P., Gelfgren, E., Hartelius, L. 2012. Communication between Huntington's Disease patients, their support persons and the dental hygienist using talking mats. Huntington's Disease – Core Concepts and Current Advances.

Ferm, U., Shalin, A., Sundin, L., Hartelius, L. 2010. Using Talking Mats to support communication in persons with Huntington's Disease. International Journal of Language and Communication Disorders.

Grimstvedt, T. N., Miller, J. U., Van Walsem, M. R., Feragen, K. J. B. 2021. Speech and language difficulties in Huntington's disease: A qualitative study of patients' and professional caregivers' experiences. International Journal of Language & Communication Disorders.

Hallberg, L., Mellgren, E., Hartelius, L., Ferm, U. 2013. Talking Mats in a discussion group for people with Huntington's disease. Disability and Rehabilitation: Assistive Technology.

Hamilton, A., Ferm, U., Heemskerk, A. W., Twiston-Davies, R., Matheson, K. Y., Simpson, S. A., Rae, D. 2012. Management of speech, language and communication difficulties in Huntington's disease. Neurodegenerative Disease Management.

Hartelius, L., Johsson, M., Rickeberg, A., Laasko, K. 2010. Communication and Huntington's disease: Qualitative interviews and focus groups with persons with Huntington's disease, family members and carers. International Journal of Language and Communication Disorders.

Hartelius, L., Carlstedt, A., Ytterberg, M., Lillvik, M., & Laakso, K. 2003. Speech disorders in mild and moderate Huntington disease: Results of dysarthria assessments of 19 individuals. Journal of Medical Speech-Language Pathology.

Power, E., Anderson, A., Togher, L. 2011. Applying the WHO ICF framework to communication assessment and goal setting in Huntington's disease: A case discussion. Journal of communication Disorders.

Rusz, J., Klempíř, J., Tykalová, T., Baborová, E., Čmejla, R., Růžička, E., Roth, J. 2014. Characteristics and occurrence of speech impairment in Huntington's disease: possible influence of antipsychotic medication. Journal of Neural Transmission.

Saldert, C., Fors, A., Ströberg, S., Hartelius, L. 2010. Comprehension of complex discourse in different stages of Huntington's disease. International Journal of Language & Communication Disorders.

Watson, K. H., Ciriegio, A. E., Pfalzer, A. C., Hale, L., Jones, M. T., Brown, B., Compas, B. E. 2021. Neurobiological and psychosocial correlates of communication between Huntington's disease patients and their offspring. The Journal of Neuropsychiatry and Clinical Neurosciences.

Zarotti, N., Simpson, J., Fletcher, I. 2017. 'I have a feeling I can't speak to anybody': A thematic analysis of communication perspectives in people with Huntington's disease. Chronic Illness.

Zinzi, P., Salmaso, D., De Grandis, R., Graziani, G., Maceroni, S., Bentivoglio, A., Zappata, P., Fontali, M., Jacopini, G. 2007. Effects of an intensive rehabilitation programme on patients with Huntington's Disease: a pilot study. Clinical Rehabilitation.

Swallowing

Bachoud-Lévi, A-C., Ferreira, J., Massart, R., Youssov, K., Rosser, A., Busse, M., Craufurd, D., Reilmann, R., De Michele, G., Rae, D., Squitieri, F., Seppi, K., Perrine, C., Scherer-Gagou, C., Audrey, O., Verny, C., Burgunder, J,M. 2019. International Guidelines for the Treatment of Huntington's Disease. Frontiers in Neurology.

British Medical Association, Royal College of Physicians. Clinically-assisted nutrition and hydration (CANH) and adults who lack the capacity to consent: Guidance for decision-making in England and Wales. 2018. London: BMA.

Hamilton, A., Heemskerk, A. W., Loucas, M., Twiston-Davies, R., Matheson K. Y., Simpson, S. A., Rae, D. 2012. Oral Feeding in Huntington's Disease: A Guideline Document for Speech and Language Therapists. Neurodegenerative Disease Management.

Kalkers, K., Schols, J. M. G. A., van Zwet, E. W., Roos, R. A. C. 2022. Dysphagia, Fear of Choking and Preventive Measures in Patients with Huntington's Disease: The Perspectives of Patients and Caregivers in Long-Term Care. Journal of nutrition, health and aging.

Keage, M., Baum, S., Pointon, L., Lau, J., Berndt, J., Hopkins, J., Maule, R., Vogel, A. P. 2020. Imaging and Clinical Data on Swallowing Function of Individuals with Huntington's Disease and Dysphagia. Journal of Huntington's Disease.

Langmore, S. E., Terpenning, M. S., Schork, A., Chen, Y., Murray, J. T., Lopatin, D., Loesche, W. J. 1998. Predictors of aspiration pneumonia: how important is dysphagia? Dysphagia.

Pizzorni, N., Pirola, F., Ciammola, A., Schindler, A. 2020. Management of dysphagia in Huntington's disease: a descriptive review. Neurological Sciences.

Rada, R. E. 2008. Comprehensive dental treatment of a patient with Huntington's disease: literature review and case report. Special Care in Dentistry.

Royal College of Speech & Language Therapists. 2021. Eating and drinking with acknowledged risks: Multidisciplinary team guidance for the shared decision-making process (adults). London: RCSLT.

Sapmaz Atalar, M., Oğuz, Özlem., Genç., G. 2024. Dysphagia in Individuals with Huntington's Disease: A Narrative Review. European Journal of Therapeutics.

Schindler, A., Pizzorni, N., Sassone, J., Nanetti, L., Castaldo, A., Poletti, B., Solca, F., Pirola, F., Lazzari, L., Stramba-Badiale, M., Rossi, A., Silani, V., Mariotti, C., Ciammola, A. 2020. Fiberoptic endoscopic evaluation of swallowing in early-to-advanced stage Huntington's disease. Scientific Reports.

Stewart, C. 2012. Dysphagia Symptoms and Treatment in Huntington's Disease: Review. Perspectives on Swallowing and Swallowing Disorders. Dysphagia.

Notes

Notes

Notes

Get in touch

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

email **info@hda.org.uk**

phone **0151 331 5444**

www.hda.org.uk

@hda_tweeting
@hdauk
@hdauk
@hda_uk
Huntington's Disease Association

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

Design and print by the Huntington's Disease Association. Published February 2026. First edition

Please note this information is correct at time of design. Visit the website to check for the most up to date information.

Inspired by our community