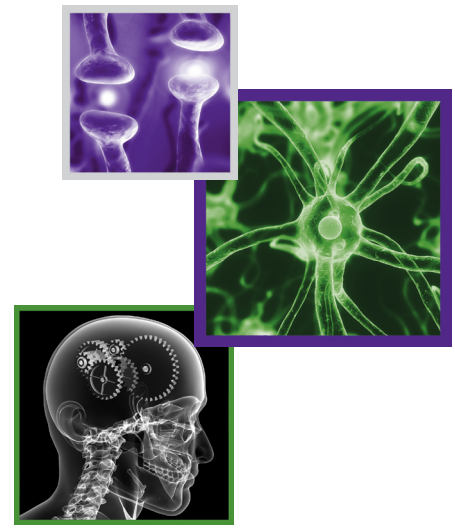
SPECIAL REPORT

Management of speech, language and communication difficulties in Huntington's disease



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Practice Points

- Speech and language therapy has an important role to play in the management of communication problems in Huntington's disease.
- As the disease progresses the effectiveness of communication becomes increasingly compromised by a combination of changes in motor function, diminishing cognitive linguistic skills and neuropsychiatric changes, such as depression and apathy.
- Signs and symptoms associated with Huntington's disease are distinctive but there is considerable variation between individuals on the extent, rate and natural course of disability within the disease. For these reasons assessment and review must be comprehensive and consideration should be given to a number of factors such as mood, motivation and behavior, which will be pertinent to performance.
- As a variety of symptoms can affect communication no single course of treatment will be effective throughout the disease. The consensus of opinion is that the therapy management will vary and that interventions will at different times be rehabilitative, facilitative, informative and supportive.
- All interventions must be timely and responsive to the changing needs of the individual and to the challenges faced by the family and others.
- Augmentative and alternative communication can compensate for communication difficulties in Huntington's disease and can increase the individual's chance for participation in daily life but strategies need to be implemented while there is still motivation and learning capacity.
- It is vital to understand the value and importance in involving family and caregivers in developing and facilitating effective communication strategies. These are well recognized for the positive impact they can have on the overall well being of the individual with Huntington's disease.

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SUMMARY Speech and language therapists play an important role in the management of communication difficulties in Huntington's disease (HD). As the disease progresses the effectiveness of communication becomes increasingly compromised by a combination of changes in motor function, diminishing cognitive linguistic abilities and neuropsychiatric changes, such as depression and apathy. The complexities and challenges presented by communication breakdown in HD require comprehensive assessment and interventions that are responsive to the changing motor, cognitive and emotional needs of the individual. The European Huntington's Disease Network Standards of Care Speech and Language Therapy Working Group has brought together expert speech and language therapists from across Europe to produce guidelines to improve the management of communication disorders for individuals with HD. The guidelines were developed with the aim of promoting timely and appropriate assessment and focused management throughout all stages of the disease. Literature was thoroughly searched and evaluated in an attempt to ensure that the guidelines are based on available evidence. However as there is a paucity of good-quality, high-level evidence the guidance is based predominantly on expert opinion and consensus. The provision of care varies widely between countries in Europe and the implementation of these guidelines aims to help improve the quality of care delivered to individuals with HD.

Huntington's disease (HD) is a hereditary, neurodegenerative disorder with progressive motor, cognitive and neuropsychiatric impairments, which can have a profound effect on the ability of the individual with HD to communicate **(Box 1)** [1,2]. It is caused by an unstable expansion of a CAG sequence within the Huntingtin, known as *HTT*, gene, which is located on chromosome 4 [3]. The protein encoded by the *HTT* gene is important in normal brain and neuronal development. The expanded CAG sequence leads to the production of an abnormal protein that causes brain cell dysfunction and ultimately neuronal cell death primarily in the basal ganglia but also the thalamus and cerebral cortex [4]. This single gene disease follows an autosomal, dominant inheritance affecting both men and women. Each offspring of an affected parent has a 50% risk of inheriting the gene mutation and developing the disease. Symptom onset usually occurs in middle age but a juvenile onset (<20 years of age) and late-age onset are also recognized.

Any of the degenerative changes associated with HD can have a profound effect on the individual to communicate.

The term dysarthria is the collective name given to the group of related speech disorders that manifest due to disturbances in muscular control. The choreiform movement disorder characteristic in HD can disrupt all systems contributing to speech production. In HD, neurodegeneration of the basal ganglia – specifically the caudate nucleus, putamen and globus pallidus [5] – results in a variable pattern of speech disturbances. These may be manifested in any or all of the respiratory, phonatory, resonatory and articulatory levels of speech with prominent effects on prosody. This type of speech disturbance is the pattern characteristic of hyperkinetic dysarthria [6,7], which is most often associated with diseases of the basal ganglia control circuit. Several studies characterizing the presence and nature of dysarthria in HD have identified a number of common features **(Box 2)** [8–11].

In the early stages of the disease the affected individual may have no symptoms or a mild or fluctuating dysarthria associated with some of the above features. Intelligibility is not likely to be reduced during early disease. With the progression of the disease, most affected individuals develop mild-to-moderate dysarthria with the choreic movements interfering increasingly with the intelligibility of speech. Hartelius *et al.* studied speech disorders in mild and moderate HD and determined that there was a significant difference in the severity of the dysarthric features between the mild and moderate presentations and concluded that the dysarthric speech signs seem to evolve in line with the general progression of the disease [11]. In the latter stages of HD, affected individuals are likely to suffer from severe dysarthria and natural speech may no longer be possible.

As the disease progresses, difficulties in executive functioning and in working memory influence the affected individual's linguistic abilities. A number of studies have investigated the characteristics of language impairment in HD and while there are some inconsistencies in the findings some common features have been identified **(Box 3)** [12–17].

In the early stages of HD, affected individuals may appear to have no deficits in language but research findings suggest some difficulties in comprehension may develop, particularly with more complex discourse [12]. The inclination to initiate conversation may also show signs of decline [13]. Consensus opinion suggests that even in the late stages of the disease when many factors have had a negative impact on language function it is thought that the affected individual will understand more than is expressed.

Some individuals may develop apraxia of speech. This adds a dimension of difficulty that may include inconsistent articulatory errors, groping oral movements and increasing errors with increasing word and phrase length. An apraxia of speech can also generate difficulties in complying with other sequenced activities, such as co-ordinating breathing and voice.

All interventions must be responsive to the changing motor, cognitive and emotional needs of the individual with HD and the challenges faced by family members and significant others. The focus in intervention is to evaluate and maximize communication skills, augment these skills whenever possible and to guide and educate carers, family members, health and social care professionals and others in the most effective ways of communicating with the person with HD.

The purpose of these guidelines is to provide information and guidance from the current evidence that is available in the literature as well as consensus views of the members of the EHDN Speech and Language Therapy Standards of Care Working Group, thereby reflecting the opinions of experts working in this field. The research base within the field of speech, language and communication intervention in HD is limited and there is a lack of good high-level evidence to support many aspects of the management of communication problems. It is hoped that these guidelines will enable speech and language therapists and others to provide optimum intervention to support the communication needs of the affected individual and his or her close partners at different stages of the disease process.

Box 1. Symptoms of Huntington's disease.

Motor symptoms
- Chorea
- Dystonia
- Rigidity
- Bradykinesia
- Gait disorder

Cognitive changes
- Difficulties planning and organizing
- Lack of initiation (activities and conversations)
- Perseveration
- Impulsivity
- Irritability and temper outburst
- Perceptual problems
- Unawareness/lack of Insight
- Difficulty in new learning

Speech, language and communication problems
- Specified in text

Neuropsychiatric/behavioral symptoms
- Depression
- Obsessive–compulsive disorder
- Irritability
- Apathy
- Anxiety

Other
- Metabolic changes
- Sleep disturbances
- Dysphagia

Methods

The development of the guidelines resulted from a number of meetings and discussions between speech and language therapists with extensive clinical expert knowledge and a special interest in the speech, language and communication difficulties of individuals with HD and their families. A literature search was performed electronically using Ovid MEDLINE (1988–2009), Ovid Embase (1988–2009) and EBSCO Cinahl (1988–2009) to identify evidence within the themes of communication, language problems and dysarthria in HD. A search strategy was developed in collaboration with the members of the Standards of Care SLT Group and search terms are outlined in **Box 4**. The literature search was updated during the development of the guideline. All relevant publications were identified and categorized to evidence statements according to the Scottish Intercollegiate Guidelines Network (SIGN 50) [101]. It is important to keep in mind that, due to the lack of scientific evidence, the present recommendations were also formulated based upon clinical experience and expert consensus from within the EHDN Standards of care SLT group. It is the wish of the group that these guidelines can indicate the gaps in knowledge and stimulate future research.

Box 2. Common features of dysarthria in Huntington's disease.

- Articulatory breakdown distorting both consonant and vowel sounds
- Inappropriate pausing within words and between words, prolonged phonemes
- Mistiming in breath control/sudden forced inspiration or expiration
- Inappropriate silences between utterances
- Harsh and strained/strangled phonation
- Hoarseness of voice
- Variability of volume control (in particular excess volume variations)

Management of communication in HD

Assessment

The timing of the onset and the progression of the various motor, neuropsychiatric and cognitive symptoms associated with HD remain uncertain especially in the early and middle stages of the disease [18,19]. Signs and symptoms of HD are distinctive but there is considerable variation between individuals on the extent, rate and natural course of disability within the disease [20]. For these reasons assessment of communication should be comprehensive and an evaluation of a number of factors pertinent to communication should be considered. Early comprehensive assessment provides a valuable baseline for the evaluation of later deficits and for the provision of appropriate support and intervention. Performance should be assessed and observed in different environments as results may vary. Pertinent factors to consider include mood, motivation and behavior, insight, memory (immediate, recent and new learning, long-term), medication, motor skills and opportunities for communication and social activities.

Assessment of dysarthria should include the following:

- Assessment of orofacial movements
- Respiratory function in speech
- Breath control and co-ordination
- Phonation
- Articulation
- Prosody
- Resonance
- Intelligibility

This is to provide a description of the speech and the musculature on which to base therapy and measure change. In all cases, the aims of the assessment should be to gather information on the parameters contributing to the dysarthria and the effects on daily living. These baseline data provide a measure of overall severity that can be reviewed and re-measured as the disease progresses.

Assessment of language skills in HD should involve a range of language assessments; both formal standardized tests and informal methods, such as checklists and observational protocols, in order to form an early hypothesis regarding the nature of the impairments and their impact. Formalized standardized assessments may not capture the early language problems in HD [12]. As with assessment of motor speech disorders these assessments should provide baseline information that underpins the therapeutic and management plans for affected individuals. Changes should be monitored, however, the timing, relevance and benefits of reassessment in a progressive condition such as HD should always be considered carefully.

Assessment should provide information on both receptive and expressive language skills and the following aspects are important to be considered [8,12–14,21]:

- Ability to process and retain information
- Ability to understand complex language
- Spontaneous speech
- Grammatical form, sentence structure and length of utterance

Box 3. Language impairments in Huntington's disease.

- Increasing difficulties in understanding complex discourse and in drawing inferences
- Latency of response
- Word finding difficulties
- Reductions in the number of words used
- Decreasing length of utterance
- Decreasing use of syntactical complexities
- Increasing susceptibility to interference
- Increasing difficulties in maintaining topic of conversation
- Problems with perseveration

- Naming/word finding
- Perseveration (words, phrases, ideas)
- Reading
- Writing

In addition to the assessment of the speech subsystems and of the changes in language integrity, information should be collated on the communication profile of the affected individual. This is the information that is relevant to the social contexts of communication as well as the skills and influences that can affect the effectiveness of the interaction. The following factors should be considered in the communication skills profile [22]:

- The communication skills of the individual, their strengths and needs
- Use of communication by the individual in their current environments
- The skills of the communication partner
- The impact of the communication difficulties on the individual and their environment, including emotional, psychological and psychosocial aspects

Communication strategies & techniques in different stages of the disease

The rate of progression and the course of the disease in HD are unpredictable and will vary significantly between individuals. A variety of symptoms can affect communication and no single course of treatment will be effective throughout the disease. The consensus of opinion is that a variety of approaches and therapeutic techniques should be considered to allow the individual with HD to maintain communication skills for as long as possible. The disease process can be over many years and maintaining communication skills from the early stages can be very important. An affected individual may be known to Speech and Language Therapy Services for up to 20 years or more. Compensatory approaches aimed at minimizing the disability and maximizing intelligibility should be considered. The goals of the therapy management will vary throughout the disease and may be informative, facilitative, supportive and at times rehabilitative. It is also likely that in the mid-to-later stages the main focus of the intervention will be with the family and/or carers both for the exchange of information and to support them in acquiring greater knowledge and responsibility for the success of communicative interactions. Training communication partners is important and should begin with careful observation and discussion about natural communication between the partners and the person with HD. Existing strengths should be encouraged and any carer who has built up a good relationship with the person with HD should be encouraged to share their ideas and also be used in any training for others as an example of good practice.

Box 4. Search terms used in the literature search.

'Huntington's disease' and 'deglutition disorders', 'pneumonia', 'aspiration', 'aerophagy', 'fluoroscopy', 'dysarthria', 'dysphagia', 'voice disorders', 'voice', 'verbal behaviour', 'communication aids', 'language disorders', 'speech disorders', 'phonetics', 'aphasia', 'vocabulary', 'semantics', 'anomia', 'articulation disorders', 'speech acoustics', 'linguistics', 'communication disorders'.

It is worth noting that recent research by Hartelius *et al.* on communicative interaction in HD indicated that family members and carers perceived some changes in communication in different ways than the affected individuals [20]. While family and carers focused on changes in speech, language comprehension, the lack of depth in conversation and need to make adjustment, the affected individuals were concerned more with the effort that it took to communicate and the amount of concentration required. It was acknowledged by all the participants in the study that there was variability and lack of initiative in communication and that there is a need for increased participation in social life in order to enhance communication.

Early stages

The consensus of opinion is early referral to Speech and Language Therapy Services is preferable. The aims of early referral are recognized as good practice in the management of long-term neurological conditions [102]. This facilitates clinical decision making on timely assessment and intervention. It also allows the individual and family members to:

- Seek and receive information and pertinent advice
- Discuss any communication difficulties
- Discuss future management and options on supporting communication
- Discuss anxieties

- To begin forming an effective therapeutic relationship with the appropriate professionals

Mild dysarthric impairments may be present in the early stages. Symptoms of dysarthria seem to evolve in line with general disease progression [11]. In the early stages a symptomatic approach to management can be used [20]. This may involve, for example, relaxation techniques and voice therapy techniques designed to normalize laryngeal tone if hard glottal attack or phonatory stenosis is identified. Stress and intonation drills to maintain prosodic features have also been found to be beneficial in the early stages [23,24].

Consensus opinion in early disease is that the merits of self awareness are important and teaching techniques for self-monitoring of speech is useful. Self-monitoring can be helpful if the rate of speech increases. It is recognized that in early disease affected individuals may rush speech in order to avoid the disruptions caused by involuntary movements. Individuals may benefit from breathing exercises co-ordinated with speech to maintain a rate consistent with physiologic capacity [24]. Mild cognitive changes may take place at this time and there may be difficulties processing complex information. It is recommended that comprehension should be assessed even in the early stages of HD [15] and that functional capacity scales may be useful at this time as these may give information on cognitive deficits that will have an effect on everyday communication.

Mid stages

Consensus opinion is that information is key to both the affected individual and the family/carers and that it is beneficial to increase contact with a partner or carer. Cognitive changes may compromise the affected individual's capacity for learning and it will become necessary for the listener to adopt special communication strategies. Problems with memory and difficulties with language comprehension and/or expression along with fewer communicative initiatives and social isolation may cause concerns and severe problems for the affected individual and their family and carers [20]. There is a need for different kinds of support and it is during this stage that advice to the family and carers about the communication environment is highly appropriate. The role of the speech and language therapist is to assess and advise on facilitating opportunities for communication in different activities and settings so that social relationships can be maintained. Affected individuals and family members often have previous experience of communication difficulties in HD and discussions involving the affected individual and family members/carers may elicit information on preferences and past experiences, which will help in planning for the future.

Late stages

Natural speech is no longer likely to be intelligible but evidence suggests some abilities to comprehend may remain. Severe motor deficits may also limit the individual's ability to write, type or directly select words or phrases from a communication device and these skills are required for many augmentative and alternative communication devices [19]. Additionally cognitive decline may limit the ability to learn and use many of the complex augmentative systems available.

Communication management is a challenge at this stage both through cognitive–linguistic impairment and severe dysarthria. In addition the affected individual may lack initiative and be apathetic towards participating in conversation. The individual may also feel frustrated and have a low tolerance towards engaging in certain activities. It is agreed that the focus should be on enhancing participation in communication activities, thereby supporting the affected individual to maintain some skills to use to maximum potential. There are some simple principles in managing communication to be observed at this stage [19]:

- Select simple systems that take advantage of previously learned skills;
- Avoid complex systems or techniques that are difficult to learn;
- Educate conversation partners well and rely on them. They can help to structure interactions and control the environment to maximum effect.

The clinical experience is that ancillary activities can coincidentally support communication skills, for example, hydrotherapy for relaxation can present a good opportunity to practise breathing and voice, music therapy for practising timing and volume control with singing, helping to maintain breathing and voice as well as word recall; taking part in activities that promote interaction, thus

limiting social isolation. Collaborative work with the multidisciplinary team can be beneficial in maximizing the opportunities for interaction and optimizing communication skills. Management plans should encourage natural speech as long as possible with supported communication techniques playing a key role. Strategies should be used consistently and should be as reliable as possible. This is particularly important in developing and establishing Yes/No systems. All communication strategies should be documented and made accessible to caregivers to limit the potential for breakdown. It is essential that the affected individual maintain some sense of autonomy and independence for as long as possible.

■ Augmentative & alternative communication

Augmentative and alternative communication (AAC) is the all embracing term for the different symbols, devices, strategies and techniques that are used to compensate for an individual's difficulties with speech, language and communication [25]. AAC interventions target the specific difficulties an individual may have in relation to both expression and understanding. More specifically, AAC includes all existing means of communication in an individual (e.g., speech and body communication) as well as different low- and high-technological communication systems and the components that are needed for these to be used in meaningful ways. Letters, words, phrases and pictures on boards and in speech generating devices and computers; digitalized and synthetic speech and different techniques and strategies for achieving effective communication are all important components of an AAC system as are conversation partner strategies.

Augmentative and alternative communication can compensate for communication difficulties in HD and increase the individual's chances for participation in daily life but need to be implemented while there is still motivation and learning capacity. It also requires significant involvement from close partners. According to the individuals with HD, family members and carers in the interview study by Hartelius *et al.* communicative initiative is a problem, indicating that support for social interaction in different daily activities is important for persons with HD and their conversation partners [20]. A recent study by Power *et al.* also discusses the importance of social interaction in the management of HD [26].

The potential for multiple complexities in presenting features including motor, cognitive–linguistic and neuropsychiatric symptoms that may interact and affect communication, are likely to affect the individual's abilities to use different communication systems. As pointed out by Beukelman *et al.* individuals with HD also seem to be underserved as far as AAC is concerned [27]; little research exists and the documentation is poor. For these reasons it may be useful for the individual clinician to make referral to or seek support from teams of professionals specialized in AAC.

Individuals with HD and their partners should be encouraged to use the 'aids' that are common and natural in many societies today, for example, paper and pen, calendars, notebooks, note lists, computers and mobile phones. The latter offer memory support functions (e.g., electronic calendars, address books and reminders) and possibilities for remote communication (e.g., Internet services, phone calls, e-mails and text-messaging [34]). With the progression of the disease these systems need to be adapted and other more specific communication devices that can be individualized may be needed. Regular assessments of communication needs and adaptations of communication systems accordingly are necessary.

In HD, visual support may be particularly important. Communication boards and books and cognitive–linguistic strategies that stimulate the use of particular words and utterances [1] can support both the understanding and production of language in the communication situation. Communication books and books with personal photographs also have been reported to stimulate communicative interaction. Visual schemes depicting different activities and their components can facilitate the planning, initiation and completion of daily activities. Personal introduction and identification cards as well as personal communication passports [28] are other low-technological resources that can fill important functions at this stage.

Communication is a collaborative project that always involves at least two individuals. There are some linguistic features that complicate communication and warrant for some particular fine-tuning and cooperation between the person with HD and his or her communication partner/s. For example, individuals with HD

have reported that other people often speak too fast and that communication rate is a factor that can influence communication negatively [20]. Family members and carers in the study by Hartelius *et al.* discussed other factors that made communication difficult for example changes in personality and physical–interactive behaviors, such as lack of eye contact. Verbal latency and echolalia are other features that can complicate communication between persons with HD and other people. Examples of advice and strategies which may support both the affected individual and the communication partner are listed below:

- Allow plenty time for communication
- Reduce environmental noise and distractions
- Talk about and set the grounds for communication, identify good and bad strategies
- How is misunderstanding signaled and resolved? What is the best communication rate? Need for pauses?
- Use of facial expression and gesture; both parties
- Use of available tools to support communication (e.g., paper and pen, picture and letter boards or other personal communication aids)
- Use a reduced rate of speech
- Repeat/rephrase and simplify message through use of key words
- Cue individual into new topics
- Use of yes/no questions
- In communication, verify understanding of the message

One method that has been shown to be successful for persons with HD and their conversation partners is Talking Mats (TM) [29]. TM is a visually based framework that helps individuals with cognitive and communicative difficulties to communicate their views. TM consists of a mat, a visual evaluation scale, pictures for conversational topics and pictures for questions relating to these. Only open questions about different issues that are relevant for the individual are asked and the person places or indicates under which picture in the visual evaluation scale a picture of a certain issue should be placed. For example, if the topic is 'health', which Hallberg *et al.* showed is relevant for persons with HD [20], and the question is "How do you sleep in the night?", the person could choose to place the picture of sleep under either of the pictures for 'excellent', 'good', 'so-so', 'not so good' or 'bad'. TM is not a personal communication aid but a resource that can be used in specific situations and with specific goals in mind (e.g., if you want to learn more about which social activities a person with HD would like to participate in). Murphy *et al.* showed that people with dementia were more effective communicators when TM was used; they became more engaged and focused in conversation and persevered less [31]. Similar findings were made in another study where TM was used by individuals in different stages of HD [32]. The affected person and their partners enjoyed using TM and felt that TM made communication easier. As it seemed the structured visual framework and the partner's interaction style facilitated the way the person with HD could think about and express their own thoughts. TM function well in face-to-face conversations involving one person with HD and one conversational partner. However, Hallberg *et al.* found that TM can also be effective in group discussions involving four persons with HD and a discussion leader [20]. Ferm *et al.* showed that both patients with HD, their accompanying significant others and the dental hygienist appreciated and felt more involved in dental and oral healthcare consultations when TM was used than when the consultations were unaided [33].

Conclusion

This guidance document on the management of speech, language and communication problems in HD has been developed reviewing the available evidence as well as expert consensus on best clinical practice. The guidelines consider the progression of the disease over time and the importance of the involvement of Speech and Language Therapy Services from an early stage in managing the complex and disruptive changes in motor function and cognitive–linguistic skills that can compromise a broad range of communication functions. It underlines the importance of responsiveness and adaptability in addressing the changing clinical picture of the affected individual and the value of involving the family and caregivers in developing and facilitating effective communication strategies. These clinical guidelines are an important tool in attempting to provide equity and quality of service provision and are also a work in progress which will require review and revision as new research becomes available.

Future perspective

These clinical guidelines mirror the fact that Huntington's disease is a complex disease that influences not only the person who has the disease but also spouses, children, friends, carers and professionals who are responsible for the care and support of affected families. Recent research and developments within the field are indicative of factors that will be important in the future; in clinical Speech and Language Therapy Services as well as in research:

- Methods for measuring changes in speech, language and communication with the progression of the disease will be developed and evaluated;
- The effects of changes in speech, language and cognition on communication will be an important issue;
- Communication and social interaction, rather than speech and language as isolated phenomena, will be of main concern in clinical practices;
- AAC systems to meet the many varying needs in persons with HD and their conversation partners will be developed and evaluated;
- Intervention programs involving the person who has the disease as well as family members and other persons in his or her social network will be developed and evaluated;
- Ecological validity and the usefulness of different intervention approaches for the persons involved – the person who has HD as well as his or her significant communication partners – should be measured and taken into consideration in clinical practice as well as research.

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■■ **This study shows that communication in individuals with Huntington's disease is sensitive to different communication modes. Communication was more effective when the picture based low technological framework Talking Mats was used than when communication was unaided. This finding is suggestive of future AAC implementation and evaluation.**

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■ **Huntington's disease leads to many different healthcare consultations in which communication is of central importance. Individuals with Huntington's disease have the right to be able to understand the information they receive from different professionals as well as to ask questions and discuss own care. This study represents an important attempt to support and evaluate communication between individuals with Huntington's disease and their carers during a naturally occurring consultation at the dental hygienist's.**

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