


Research Report

Speech and language difficulties in Huntington's disease: A qualitative study of patients' and professional caregivers' experiences

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Abstract

Background: Huntington's disease (HD) is a neurodegenerative disease characterized by a triad of motor, cognitive and psychological symptoms, leading to a gradual breakdown of communication skills. Few studies have investigated how people affected by HD and their professional caregivers, for example, medical doctors, physiotherapists and nurses, experience the patients' gradual loss of speech and language.

Aims: To examine communication-related experiences of patients and professional caregivers. Experiences with speech therapy and the use of augmentative and alternative communication aids (AAC) were also investigated.

Methods & Procedures: Seven individuals with HD and seven professional caregivers were interviewed individually, using a semi-structured interview guide. Transcripts were analysed using a conventional content analysis, and the results presented in three main categories.

Outcomes & Results: Most individuals with HD were aware of having communication difficulties, struggling with understanding others as well as being understood. This was confirmed by professional caregivers, who also raised ethical issues encountered when patients struggled with communication. Both groups talked about external factors (such as noise or crowded social settings) as disrupting communication, and shared recommendations on how people in general, and speech and language therapists (SLTs) in particular, could optimize communication. Very few patients had received information about communication aids, and none was using AACs. Professional caregivers underlined the importance of interdisciplinary collaborations, including SLTs, in order to optimize care.

Conclusions & Implications: Findings shed a light on everyday communication challenges faced by people with HD and their professional caregivers, and the lack of implementation of communication aids in this group. The dramatic impact of HD on patients' communication skills underscores the need to include SLTs in the follow-up of this patient group, ideally from the early stages of the disease, while the patient is still capable of voicing his/her own wishes and thoughts. Future research that explores how to optimize communication and implement the use of AACs for individuals with HD is needed.

Keywords: Huntington's disease, communication, language, speech therapy, augmentative and alternative communication (AAC), qualitative.

What this paper adds

What is already known on this subject

- Although the ability to communicate gradually deteriorates in individuals affected by Huntington's disease (HD), there is little knowledge about how affected individuals experience the loss of speech and

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language skills. Interdisciplinary care is recognised as essential for this patient group. However, professional caregivers' thoughts and experiences of communicating with their patients have not been fully explored. Recent years have seen a rapid growth of available communication supporting technologies that could potentially be helpful for individuals with HD, but limited attention has been given to this subject.

What this paper adds to existing knowledge

- What do we now know as a result of this study that we did not know before the results highlight that patients are aware of problems with speech and language even in early phases of the disease, and include patients' personal outlook on problems with communication. Professional caregivers raised ethical issues encountered when patients struggled with communication. Both groups described specific strategies that could facilitate communication. There was a significant lack of experience and knowledge about augmentative and alternative communication aids (AACs).

What are the potential or actual clinical implications of this work?

- Clinical implications of this study. The introduction of communication aids in the follow-up of patients with HD needs to be discussed with the patients in the early phases of the disease, and implemented while the individual still has the capacity to learn and take advantage of alternative communication support. Results underline the importance of including speech and language therapists (SLTs) into multidisciplinary care of patients with HD. SLTs should be available also for professional caregivers who need advice on how to facilitate conversations and social interactions, in order to optimise care of patients with HD.

Introduction

Huntington's disease (HD) is an inherited neurodegenerative brain disease with a 50% risk of genetic transmission (Roos 2010). The occurrence of HD is 5–14 persons per 100,000 (Baig *et al.* 2016). Symptom onset usually occurs between the third and fifth decade of life, affecting physical, psychological and cognitive functioning, and death within 15–30 years (Novak and Tabrizi 2011). More specifically, HD is characterized by a triad of symptoms, including motor symptoms, such as involuntary movements and impairment of voluntary movements, cognitive deterioration and psychiatric and/or psychological symptoms, such as irritability, depression, anxiety and apathy (McColgan and Tabrizi 2018).

HD is currently described as a multisystem neurodegenerative disease, characterized by a progressive atrophy and thinning of the cortical mantles in all four cerebral lobes (Rüb *et al.* 2016), including not only the striatum but also the cerebral neo- and allocortex, thalamus, pallidum, brainstem and cerebellum (Rüb *et al.* 2016). Language and speech are complex processes, and motor, cognitive and emotional function will, independently and in interaction, influence the ability to communicate. Speech production also requires cooperation between the nervous system and approximately 100 different muscles (Godefroy 2013).

Any of the degenerative changes associated with HD will gradually interfere with speech and language skills, and affect basic processes involved in communication and social exchange (Hamilton *et al.* 2012,

Saldert *et al.* 2010, Zarotti *et al.* 2019). Memory and attention problems, reduced processing speed and loss of cognitive efficiency are known to cause subtle changes in speech and communication already in the early stages of HD (Hamilton *et al.* 2012). More specifically, motor impairments associated with HD may affect respiration, phonation, timing and articulation (Hartelius *et al.* 2010). Most people with HD will also develop dysarthria, such as changes in cadence, tone of voice and loudness, leading to detrimental effects on the individual's speech (Hartelius *et al.* 2010). Following a decrease in cognitive function, language components will also suffer, with word-finding problems, reduced vocabulary, difficulties with speech production, understanding and semantic processing, decreased syntactic complexity, and delay in processing of spoken language, understanding and replying (Gagnon *et al.* 2018, Hamilton *et al.* 2012, Hartelius *et al.* 2010, Johnson and Paulsen. 2014, Saldert *et al.* 2010). A lack of initiative in social settings (Gagnon *et al.* 2018), in addition to reduced mimicry, and non-verbal communication being affected by the movement disorders, will also impact on communication (Pollard 2008). After a while, communicating through writing will also be difficult as a result of reduced muscle control and movement disorders. During the last phases of the disease, some people will lose the ability to speak completely (Power *et al.* 2011).

Communication is of central importance for all human beings, and communication difficulties may therefore lead to frustration, anger, sadness or depression, and have a deep impact on quality of life (Galts

et al. 2019, Neumann *et al.* 2019). There is currently no cure for the disease (Roos 2010), and clinical experience and research confirms that the emotional burden is high for all involved parties: affected individuals, relatives and professional caregivers such as medical doctors, physiotherapists and nurses (Dale and van Duijn 2015, Domaradzki 2015). In spite of the many speech and language difficulties, people affected by HD express a need for interaction with others (Hartelius *et al.* 2010). Therefore, there is a need for research exploring how communication can be optimized and facilitated in this patient group, and whether the provision of communication aids could reduce the emotional burden of communication difficulties in HD.

Augmentative and alternative communication tools (AAC) can constitute an addition or an alternative to natural speech. Nevertheless, there is a paucity of research evidence to underpin recommendations for people with communication difficulties (Baxter *et al.* 2012), in particular in neurological degenerative conditions such as HD (Beukelman and Ball 2002). The management of speech and language difficulties in HD must take the progressive nature of the disease into account, should be individually tailored, and intervention strategies should be simple to understand and use. According to the literature, and supported by clinical experience, few individuals with HD are given the opportunity to use such interventions (Ferm *et al.* 2010). We also know that support from professional caregivers is needed to help people with communication difficulties, such as in HD, exploit the potential that AACs bring (Baxter *et al.* 2012). This is a challenge to speech and language therapists (SLTs), as these technologies should be integrated into general SLT services beyond that of specialist AAC provision. Given the inherent nature of speech and communication difficulties in HD, there is a need for more knowledge about whether and how AACs could support individuals with HD.

Research has explored how HD affects speech and language and the affected person's ability to communicate (for a review, see Gagnon *et al.* 2018), and the European Huntington's Disease Network (EHDN) Standards of Care Speech and Language Therapy Working Group has produced guidelines to improve the management of communication disorders for individuals affected by HD (Hamilton *et al.* 2012). Nevertheless, clinical experience suggests that non-specialized professional caregivers lack knowledge about how to facilitate communication in this group. Also a few studies qualitatively investigate how affected individuals experience the loss of communication skills (Hartelius *et al.* 2010, Zarotti *et al.* 2019). Furthermore, little research has been conducted on how professional caregivers and SLTs may facilitate communication in this group, whether AACs could support people with HD, or how to implement such interventions in the patients'

daily life (Ferm *et al.* 2010). Professional caregivers' perspectives are also important for research aiming at improving practice. Given this gap in the literature, the aims of the current study were to examine: (1) how individuals with HD and professional caregivers in institutions providing care to patients with HD experience communication, speech and language problems associated with the disease; and (2) investigate both groups' experiences of collaboration with SLTs, in addition to thoughts about and experiences with AACs.

Methods

Research approach

The study was conducted as part of a master's thesis and a research project at the Centre for Rare Disorders, Oslo University Hospital, Norway. The methodological approach was a conventional content analysis (Hsieh and Shannon 2005). This approach was initially developed to deal with the objective and manifest content of data, and was therefore deemed to be a suitable choice for the analysis of the current data, given potential restricted speech and language skills in participating patients. The same method of analysis was used for the professional caregivers, so that results could be presented with a similar structure. Content analysis is useful when knowledge and previous literature on a phenomenon is limited, with a systematic classification process of coding and identification of categories or patterns (Hsieh and Shannon 2005). Content analysis provides the opportunity to emphasize individual variations, and offers the possibilities of using manifest and descriptive content (visible and obvious components), as well as latent and interpretative content (interpretation of underlying meaning) (Graneheim *et al.* 2017, Graneheim and Lundman 2004).

Setting

The Centre for Rare Disorders is a national resource centre responsible for developing and transferring knowledge about rare conditions, such as HD. The country is also organized with five regional resource centres for patients with HD, providing help and support to patients in later phases of the disease. Several different professions are involved in the care of patients with HD, for example, medical doctors, physiotherapists and nurses. Some of these centres have daycare support units and residential care/nursing homes. In addition, there are two rehabilitation centres for patients with HD.

Participants

Patients were informed about the study and recruited when on rehabilitation stays or in daycare centres in

Huntington institutions in eastern Norway. Professional caregivers were recruited from the same institutions. Recruitment was mainly conducted by J.U.M., who at the time of the study worked at the Centre for Rare Disorders and at an institution with a separate department for patients with HD. Potential participants with HD, seven in total, were informed about the study. All seven consented in participating. Information was provided in written form, but also orally for patients who wanted this, in order to secure understanding of the study and what participation would entail. Participants provided written consent, and were informed that participation was voluntary and that they could withdraw at any time. Interviews were performed between November 2018 and January 2019 at one of the two institutions where patients and professional caregivers had their connections.

The study included 14 participants: seven patients in the early to middle phases of HD (Group 1) and seven professional caregivers of patients with HD (Group 2). Six of the patients attended a daycare centre/rehabilitation centre, whereas one patient lived in a care home. There were four males and three females. Four patients were between 18 and 35 years old at the time of the interview, whereas three were aged between 35 and 65 years. Two patients had been diagnosed with HD less than 5 years ago, and five patients had been diagnosed for more than 5 years. Information about when HD had been diagnosed was provided by the patients themselves. In Group 2, four professional caregivers represented different healthcare professions. Group 2 also included three SLTs. Most professional caregivers had experience with patients in the middle to late phases of HD. There were five females and two males among the professional caregivers.

Inclusion criteria for Group 1 was having HD, with or without language and/or speech difficulties, and be willing and able to participate. Hence, adjusted convenience sampling was applied, and individuals in contact with the research team during the recruitment phase were approached. The evaluation of whether patients were relevant for participation was done by J.U.M., based on the potential participant's communication skills in a daily care setting. Following general principles of research ethics, we wanted to ensure that patients had the physical and cognitive capability to share their experiences, understand the meaning of the questions asked and for the interviewer to understand the answers they provided. People with moderate to severe speech and/or language difficulties who would not be able to express themselves in an interview situation were therefore not informed about the study and would have been excluded.

The only inclusion criterion for Group 2 was having work experience with people with HD. We wanted

professional caregivers to represent different professions, in order to provide different care perspectives, and this was achieved. Participants from Group 2 were working at the same institutions as potential participants, but were not necessarily involved in the care of participating patients. Participant from both groups were recruited from the same institutions for practical reasons, but also to secure similar reference frames.

Procedures

Semi-structured interview guides were developed, one for patients, and one for professional caregivers, based on one of the authors' (J.U.M.) longstanding clinical experience with patients with HD, and the current literature. Questions focused on the areas of communication, experience with speech therapists and communication aids. We wanted questions to Group 1 to be clear, concise and easy to understand, while at the same time allowing patients to share their personal experiences. Pilot interviews were performed by T.N.G. on fellow students and subsequently revised as a result. Both interview guides consisted of 16 main questions (see appendices A and B).

Semi-structured in-depth interviews were conducted face to face by T.N.G., were audio-recorded, transcribed verbatim using HyperTRANSCRIBE and anonymized. Interviews were chosen as a method since it allows for a flexible approach to subjective experiences, and is especially useful when the aim of a study is to produce a better understanding of subjective experiences. Three SLTs, who contacted T.N.G. after hearing about the study, were included after completion of the oral interviews, and provided additional data. Resources were at the time not available for in-depth interviews of the three SLTs, and these participants therefore completed the interview guide in a written form.

Data analysis

Analysis was performed using the procedure described by Hsieh and Shannon (2005). Only overt statements, coming directly from the participants, were included, whereas questions from the interviewer answered only by yes or no were left out of the analysis, since we could not be certain that the participant had understood the question correctly.

T.N.G. performed a preliminary analysis of the data for her master's thesis. Given the length and structure of the thesis, the authors decided to reanalyse the data for the purpose of the present paper, so that the results could be synthesized and presented in more precisely. Therefore, the original transcripts were reanalysed and recategorized according to code similarity. During the

Table 1. Examples of coding

	Code
<i>Text extract (participants with HD)</i>	
[W]hen people talk. And they talk softly. With a lot of sound all around, then ... then it becomes very little. I get out of it because therefore (pause) then you can't all the time ask 'what did you say' because that is difficult too. Understand all that goes on.	Noise (disturbs communication) Understanding
[B]ecause when I am, no, because I speak worse and then I notice that they are many they because. They have, they do not take their time. Or, to, in a way listen enough. Because when I say something or so, then often I try.	Patience (listener)
'It can be things like that. Worse and worse ... to talk, yes. To communicate.	Disease progression
'It is much easier with someone you kind of knows what it is all about, whereas others ... don't have a clue. And then it is so <i>much</i> it affects and, so much to explain and yes. Things ... that belongs to the disease.	Knowledge about HD (advantage)
<i>Text extract (professional caregivers)</i>	
Some patients have lost all speech. And some have very very limited language skills. So we have days when we have to ... almost guess. So we base our [guesses] on how well we know the patient. We have some knowledge about the patient's interests, what they usually ask and things like that. But it may be almost impossible to really understand what the patients mean.	Understanding Guessing Knowledge about the patient
Sometimes, understanding what they say is completely impossible. And if they experience that we do not understand them ... they become very stressed and talking is even more difficult. Sometimes, a time-out is necessary, and you come back after 10 minutes, and you try again.	Understanding Strategies to enhance communication

second phase of analysis, J.U.M. and K.B.F. read all the interviews several times to obtain a sense of data content. After having read all interviews several times, the same two authors separately read the transcripts word by word while also deriving codes. Words and sentences that appeared to capture key thoughts or concepts were highlighted. During this process, labels for codes emerged that were reflective of the content of given categories, which were used to organize the data into meaningful clusters (subcategories). Codes and categories were subsequently compared with results presented in the master's thesis, and results were similar. Examples of coding for both groups can be found in table 1.

Credibility and trustworthiness

Trustworthiness is about making it clear whose voice, the participants' or the researchers', is heard in a study (Graneheim *et al.* 2017). This was secured by using representative quotes from the transcribed text, and credibility was sought by achieving agreement between all co-authors (Graneheim and Lundman 2004). To increase rigour and trustworthiness, the two first authors and the last author read all transcripts in detail, and subsequently examined and discussed codes and categorization of the data, to ensure the reasonableness of the analysis. The participants were given the opportunity to listen to the recorded interviews and read through the transcriptions in order to provide va-

lidity checks, but none of the participants used this opportunity.

Ethics

The study was submitted to the Regional Committee for Medical Research Ethics (REK), which concluded that the study could be carried out without its approval. The study was therefore described and recommended by the Data Protection Officer at Oslo University Hospital (18/11423).

Results

Categories can be used in content analysis when describing the data on a manifest level (Graneheim *et al.* 2017). Data were structured within three central categories, reflecting the main issues covered by the interview guides: communication (category 1), experience with speech therapy (category 2) and communication aids (category 3). The first category includes three subcategories. The categories, subcategories and results are presented and summarized in table 2 and will be described in more detail below. Subcategories differed slightly between the two groups of participants. To enhance structure, the results will be presented separately for the two groups within each category. Categories and subcategories are supported by direct exemplar quotations from participants, who have been given pseudonyms to preserve their anonymity.

Table 2. Overview of the categories and subcategories, and summary of the results for both participant groups

Patients	Professional caregivers
<i>1. Communication</i>	
Talking to people: an activity they appreciate or love Importance of time, listening, and empathy	Convey a message Value of communication Consequences of lost communication skills
<i>1.1 Impact of HD on speech and language skills</i>	
Factors experienced as disturbing: <ul style="list-style-type: none"> • Noise (many people, external noise) • Cognitive aspects (processing, memory, and attention) • Speech impairment (sound, clarity, airflow) • Communication partner (interrupting, lack of social inclusion, lack of patience) 	Variation across patients and disease phases Speech and language impairments (quantity, speed, quality, relevance) Body language (mimic, gestures) Cognitive aspects (meaning and content) Interpretations and guessing needed in later phases
<i>1.2 Optimising communication quality</i>	
The importance of: <ul style="list-style-type: none"> • Experienced trust • Avoidance of prejudice and generalising • Positive outlook, understanding, empathy, caring approach, patience • Knowledge about HD 	Skills and strategies: <ul style="list-style-type: none"> • The ability to read and use body language • Support from people who know the patient well (colleagues, family, friends) • Clear and concise language • Reduce noise and distractions • Usefulness of routines, time and patience
<i>1.3 Disease progression and changes</i>	
Aware of changes, but struggle with specifically describing them Aware of changes in cognitive abilities: concentration, memory, and multitasking.	Describe progressive changes affecting communication and conversational content Later phases characterised by mostly practical and physical needs (hunger, pain)
<i>2. Experiences with speech therapy</i>	
Most patients had regular follow-up from SLTs and described it as helpful: <ul style="list-style-type: none"> • Importance of using simple language, limit amount of information, time, patience, and knowledge about HD • Training of cognitive, speech, and language skills • Singing particularly enjoyable • SLT group sessions also mentioned as important, enabling them to meet others in a similar situation 	Non SLT professional caregivers experienced communication and speech difficulties as too complex for speech therapy to give results SLTs highlighted the importance of multidisciplinary teamwork including a focus on communication SLTs could guide professional caregivers in order to maximise patient support
<i>3. Experience and thoughts about communication aids</i>	
Most had received information, but knew little about possibilities and options One patient had plans of using AAC One patient had some but limited experience with AAC	Importance of early and preventative introduction of AACs when patients are still able to learn to use the aid Most professional caregivers had no experience and little knowledge about AACs for patients with HD

Translations of quotations used to illustrate the results attempted as much as possible to be true to the exact wording used by the participants with HD. Hence, errors of language were kept. Brackets [] are used to clarify meaning, such as when the participants refer to previous information. Parentheses (...) indicate that some text has been removed.

Category 1: Communication

All participants with HD described communication as talking to people, and three specified that communication was something they appreciated or loved. A few patients mentioned characteristics they felt were important for communication, such as time, the communication partner's ability to listen and empathy.

That people have time to talk to me. (...) It is GP [general practitioner]. They are very understanding. (...) Talks about how I feel the whole person. And I experience also now from you that you care about. Whole me. And I appreciate that. Yes, yes, it is like that. Fellow humans make my everyday life easier. (Victoria)

Professional caregivers explained the meaning of communication in a more academic way, talking about conveying a message and the value of understanding each other. Professional caregivers also reflected upon the emotional consequences of losing the ability to communicate.

They have this strong wish to communicate, that they used to manage. But their tongue kind of curls up, and they can't express themselves. They become frustrated

and stressed, and we become frustrated and stressed, and it all turns ... chaotic. Sometimes. (Sylvia)

Subcategory 1: The impact of HD on speech and language skills

Group 1: Participants with HD

When asked about aspects of communication that could be challenging for them, two participants stated that they had few problems with communication, whereas five experienced difficulties with understanding and talking. When asked in more detail, participants mentioned factors that could disturb communication, such as speech difficulties, or external factors such as noise and disruptions. Responses varied across participants, and most factors were mentioned by only a few, except noise that was raised as an issue by five participants. Conversations with several people were described as noisy and difficult to follow, as well as situations involving external interfering sounds, such as public transport.

The worst is when I ... and someone talk. And they talk lower. Or someone talks. With lots of noise around, then, it is very little. I become more lost, because you can't ask what they said all the time, because that is what is difficult to do as well. Process things. (Sophie)

Two participants mentioned the challenge of meeting new people, and three participants felt that understanding and processing new information was difficult. Memory problems were mentioned as a challenge by one participant, whereas two raised the issue of difficulties with concentration and multitasking. Another participant talked about fears of not being able to understand expectations or written demands coming from social services.

Six of the seven participants felt that they still managed to express themselves and be understood by others, in spite of increasing difficulties. Nevertheless, three had experienced that others could struggle with understanding their speech, and two participants explicitly mentioned speech difficulties as disrupting communication. When asked about which aspects of speech they experienced as difficult, participants struggled with being more specific, but mentioned challenges with air-flow or breathing, lack of clarity, and difficulties with the voice (sound). Two participants talked about other people's lack of patience, and experiences of being interrupted by others, or not being included in other people's conversations.

It is ok with the physiotherapist here, because it is only the two of us. But I notice that if I am invited at a party, it is easy to end up just sitting there in the corner, not

saying much, yes. That it becomes a little too much. Talk from the others. (Mary)

Group 2: Professional caregivers

Four of the caregivers specifically highlighted the large variation in communication skills across individuals. The disease was described as affecting the patients' ability to talk (speech), language skills, use of gestures and mimic, body language, and cognitive understanding of meaning and content. The three SLTs mentioned that with disease progression, patients talked less, speech was slower and more indistinct, answers were less relevant or the same questions were asked several times. Differences were primarily described as related to disease phases, but one professional caregiver also described how she had observed variations across individuals, irrespective of disease phase.

Two of the professional caregivers talked about the discrepancy between the patients' intention of what was being communicated, and the recipients' understanding of it. This was explained by the rapid progression of the disease in some patients, by a lack of awareness of disease status, or due to physical HD-related impairments.

Speech is unclear. So ... sometimes it looks as if the patient believes that what is said sounds reasonable. While for us ... it sounds like muttering. (Paul)

In later phases of the disease, caregivers not only described difficulties understanding what the patients said, but also difficulties knowing whether messages were or were not understood by the patients. HD was described as mitigating facial expressions and gestures that normally could have supported the gradual loss of speech and language. In addition, patients' feelings were described as less obvious, more difficult to read or taken away by the disease, complicating the caregiver's interpretation of the patients' needs or understanding. As a result, caregivers had to guess when communicating.

We end up having to decide things over their heads, because we assume we won't even be able to create a dialogue that could help us understand what they really want. (...) Based on their reaction, it may be difficult to interpret whether they have understood or not. Usually, you can see if people are in pain. When you can't even [communicate that you are in pain], we have a problem. (Anthony)

Subcategory 2: Optimizing communication quality

Group 1: Participants with HD

Three participants shared their thoughts about how other people could strengthen and improve the quality of communication when talking to them. The

importance of experienced trust was mentioned by one. One participant also talked about the importance of other people not generalizing or being prejudiced. Participant also described what they saw as important characteristics for an optimal dialogue, such as a positive outlook, understanding, empathy, being seen as a whole person and caring. Patience was mentioned by two participants as crucial, and one of the participants explicitly described how patience from close family contributed to feelings of having a good relationship with people she cared about.

It is important that they have enough time. And that they do not interrupt right away and or talk about something else. (Sophie)

When asked, four participants confirmed that other people's knowledge about HD could be important to improve the quality of communication, since it would lead to a better understanding of encountered difficulties, and a more correct interpretation of symptoms.

Group 2: Professional caregivers

Caregivers were asked how they knew whether they had understood the patient. While one of the professional caregivers seemed to have given up on trying when speech was lost, most caregivers had developed skills and strategies that could help them improve communication, in spite of the many challenges with speech and language. As an example, caregivers did their best to read the patient's body language, exaggerated their own facial expressions and gestures, and/or used colleagues who knew the patient well to increase understanding.

[The patient] becomes stressed. Or his/her gaze flickers, like looking around searching for someone that could help and explain. Turn like, not irritated, but giving up in a way. (Jane)

When communication had been reduced to guessing because of the progression of HD, professional caregivers experienced the worth of knowing the patient well, or using someone well acquainted to the patient, such as family members or friends, in an attempt to counteract the lack of communication skills. Knowledge about HD was also described as central when trying to understand or guess what the patient was communicating.

Maybe it becomes easier, when I have got to know them. (...) Which helps me, I guess more easily, because I know them. Lots of guessing. Lots of 'this?', 'this?', 'or this?', and then you see it when you have guessed correctly. (Jane)

Professional caregivers had experienced how communication could be improved by using short and simple sentences, talking slowly, and using precise language. Quiet places the patient knew well and felt safe in were also described as important. Routines were also considered central. Caregivers knew that external stimuli and noise could be disrupting, and face-to-face settings with minimal disturbances and focused activities were therefore chosen, if possible. In addition, time and patience were described as imperative, as well as letting the patient chose topics of conversations. Professional caregivers had also experienced that understanding could be complicated by introducing a change of topic. The caregiver's persistence and ability to find alternative solutions when communication was severely impacted by the disease also seemed crucial.

Subcategory 3: Disease progression and changes

Group 1: Participants with HD

Participants with HD were asked about whether they felt their communication skills had changed over time, after being diagnosed with HD. Five participants answered that there had been some changes, but struggled with being specific when asked. When managing to describe changes they were aware of, participants talked about increasing difficulties with speech and communication, concentration, multitasking and memory.

I have always been ... no. I have more difficulties with speech today. As you can hear. Yes. But I like talking about the disease. (Victoria)

One participant talked about the relief of receiving an explanation (diagnosis) for her increasing difficulties, and tried to approach the disease accepting its reality, while another participant felt that time had helped him feel more confident in spite of the many difficulties with communication.

Group 2: Professional caregivers

Professional caregivers described how disease progression could be sudden, dramatically affecting the quality of communication between patients and caregivers. One participant shared the emotional burden she felt when the health of patients she knew well worsened, and ways to communicate were lost.

The content of conversations was expectedly affected by the progression of the disease. Hence, in later phases of HD, communication was mainly restricted to practical and physical needs, such as hunger or pain.

I have followed one patient in particular for a long time. I have observed a huge gap [in the patient's condition]. We had really good communication. Talked a

lot with each other. Now ... well, we do not talk at all.
(Paul)

Category 2: Experiences with speech therapy

Group 1: Participants with HD

All patients except one had regular follow-ups by a SLT, weekly according to three participants. One of the participants did not feel he needed any help with speech or language, although the interview data show that also this participant's speech was affected by HD.

Five of the seven patients described speech therapy as including exercises that would increase speech volume, whereas three participants mentioned cognitive and language training. Singing was also described as part of training by three, an activity all three specifically described as enjoyable. Relaxation and tongue exercises were mentioned by one participant. Four participants felt that speech therapy was useful and helped them progress.

When asked about what SLTs could do to strengthen communication, one participant talked about the importance of using simple language and few words, and limit the amount of information. Another participant talked about time and patience, and having relevant and necessary knowledge about HD. Two of the participants stated that the crucial factor for positive communication was having normal conversations, and the experience of professional caregivers caring about how they coped with the disease.

There is not much ... not very much knowledge. Because [the SLT] has learned [about HD] by himself, joining some courses. (...). So he only has some general background. (...) Like I said ... maybe be more ... yes, a little bit more concerned about how I actually have ... with the disease. (John)

Four participants also participated in Huntington-group sessions with a SLT, an activity that was described as particularly enjoyable by two, since it gave them the opportunity to meet others with HD, and learn group-wise.

Group 2: Professional caregivers

All four professional caregivers who were not SLTs were or had been working with patients in the later phases of HD, and described communication difficulties in the late phases as too complex for speech therapy to be really efficient. Three of the four caregivers (non-SLTs) had not received any training in how to improve communication in patients with HD. One of the caregivers had received training, but described it as not very useful.

As I see it, the disease has progressed too much. So speech therapy is not very efficient by then. When we started offering speech therapy to our residents [some time ago], many of our patients were on a completely different level. At that time, speech therapy was really useful. And they managed to practice. (...) But now I feel ... I feel they don't really get anything out of it. (...) In theory this is probably very good, but it becomes really difficult for them. They struggle with keeping focus and ... yes, this is so much more complex. (Paul)

Two of three SLTs described the importance of multidisciplinary teamwork in the follow-up of patients with HD, in order to maximize support.

Multidisciplinary collaboration is of high priority in our practice. Which means that all professional caregivers who work with patients with HD can share strengths and weaknesses, and the SLT can provide information about assessments and give advice to promote good communication. (Emily)

The usefulness of an interdisciplinary approach with support from SLTs with specific knowledge and experience with HD was also mentioned by some of the other caregivers.

Communication is a huge part of HD. So there should clearly be more focus on it. (...) I believe SLTs should become more included in treatment. Or part of a patient's follow-up. (...) Absolutely. Regular meetings where we could have discussed professional challenges. (Paul)

Category 3: Experience and thoughts about communication aids

Group 1: Participants with HD

Five participants had received some information about alternative communication aids, whereas two had not. Three participants used their phone as support in daily life, mainly as a memory aid. In spite of having received information about potential communication aids, only one participant was planning to try a specific communication aid in the near future, and one had some experience with using it. However, none of the participants managed to describe how communication aids could be or had been helpful. The most specific descriptions were given by the participant having some experience with a communication aid.

The speech therapist visits me, now. So we work with understanding this communication aid. So she will come one day. To me. (...) She came. Was to order it, then so. (...) I save everything on all different I want to have. Exactly this. (...) I can for me to perhaps be able to communicate with people. I can save you on a ... there then. Because talking becomes more and more

difficult. And then, yes, I can save it on the [communication aid]. (Matthew)

Group 2: Professional caregivers

An early and preventative introduction of communication aids was mentioned as important. However, only one of the seven caregivers had actually used communication aids with patients with HD, and explained this with a lack of knowledge about existing aids, and an impression that extra time would be needed if they were to communicate alternatively. As illustrated by the following quote, introducing communication aids earlier than needed could also stir negative reactions in patients who felt they did not need communication support.

I believe that all patients with HD would benefit from communication aids. However, I feel [aids] are introduced too late. Such aids should be introduced from the beginning, so that they manage to learn how to use it. The problem, however, is that at that stage, they don't want to, since they are not sick. So when they reach the phase when they are sick enough to need it and they would like to learn, it doesn't work optimally. This is so complicated. (Paul)

Two caregivers emphasized the importance of using communication aids that were easy to use and simple to understand. Books with drawings and pictures of objects, people or activities, were described as facilitating conversations for patients with restricted communication skills. However, caregivers pinpointed the limitation of such communication aids when a patient or professional caregiver needed a conversation beyond simple pointing.

Discussion

The aim of the present qualitative study was to explore how individuals with HD and professional caregivers experienced speech and language problems associated with the disease, their experience with speech and language therapy, and their thoughts about and experience with AACs. Results highlight that individuals with HD were aware of having communication difficulties, and some managed to describe specific challenges and situations they could experience as difficult. Professional caregivers confirmed speech and language difficulties in this group, and raised ethical issues encountered when patients struggled with communication, such as arising dilemmas regarding care options and choices when professional caregivers did not understand the patients' wishes or needs. Both groups provided specific recommendations for how people in general, and SLTs in particular, could optimize communication. Very few of the patients had received information about communica-

tion aids, and only one had some but very limited experience with using an AAC at the time of the study. Findings can help shed light on everyday communication challenges faced by people with HD and by their professional caregivers, and have implications for clinical practice and future research.

The experience of communication difficulties in HD

Patients and professional caregivers addressed a range of disease characteristics that led to challenges in communication, in line with previous research and described in guidelines (Gagnon *et al.* 2018, Hamilton *et al.* 2012). Interestingly, although the interviews confirmed difficulties with language processing and expression, patients as a group found it easier to describe difficulties related to speech. This is in line with Hartelius *et al.* (2010), who found that affected individuals were more concerned with the effort involved and concentration needed in speech, whereas relatives and carers focused on comprehension, lack of depth in conversations and the need to make adjustments. On an individual level, however, some participants were aware of problems with understanding, processing, memory and concentration.

Patients and professional caregivers underscored the importance of patience and time. Consistent with the literature (Hamilton *et al.* 2012), some participating patients had long latency of response, used simple language, distorted syntax or vocabulary, and had difficulties finding words. Hence, communicating with individuals affected by HD requires time, which may be a challenge for busy professional caregivers. Professional caregivers also highlighted the usefulness of interpreting the patient's body language. However, symptoms of HD, like chorea, reduced mimicry and apathy, are well known to affect non-verbal conversational elements, further exacerbating communication difficulties.

Both groups described the disturbing effect of noise, disruptions or cognitive symptoms as complicating communication, as also described in HD guidelines (Hamilton *et al.* 2012). Communication barriers had a social impact and led to feelings of social isolation. This finding clearly illustrates the potential association between speech and language difficulties and quality of life, and underlines the importance of optimizing affected individual's opportunities for communication, in order to promote social inclusion and engagement, thereby reducing the emotional and psychological burden of HD as much as possible (Hartelius *et al.* 2010).

Optimizing communication

As the disease progresses and compromises communication, approaches aimed at facilitating conversations

need to be individually tailored and take disease progression into account (Hamilton *et al.* 2012). The importance of creating optimal communication environments where external disruptions, if possible, are reduced to a minimum, need to be taken into account in clinical settings.

When struggling with communication, professional caregivers also highlighted the importance of involving people that knew the patient well, as demonstrated in individuals with Parkinson's disease (Johansson *et al.* 2020). This could be family members, but also other professional caregivers who know the patient well. In an effort to counteract some of the barriers of communication in HD, relatives could provide information, pictures, and a history of the patient before HD developed, information that would support professional caregivers in understanding the patient, personalize care and improve communication (Handberg and Voss 2018). Our findings also shed light on the importance of knowledge about HD and its associated difficulties, knowledge that may increase the chances of the other person being able to facilitate a conversation and adjust to challenging communication difficulties. Involving people who know the patient well, and facilitating knowledge about HD, is in line with the recommendations from the EHDN Standards of care SLT group (Hamilton *et al.* 2012), who also recommend early referrals to SLTs in order to initiate a therapeutic relationship. Getting to know the patient before speech and language difficulties severely impedes communication, may support the professional caregiver's ability to guess what is being said or wanted, and may strengthen feelings of trust, which in turn may reduce levels of stress in patients (Zarotti *et al.* 2019).

Including SLTs in the follow-up of patients with HD

Early referrals to SLT services are important to minimize the progressive adverse effects of communication loss (Hamilton *et al.* 2012), and case studies show that speech therapy may help to reduce the progression of communication difficulties in individuals with HD (Bilney *et al.* 2003). In the present study, four of seven patients felt that speech therapy was useful, and patients also mentioned the positive impact and subjective value of HD group sessions run by a SLT. Although the objective value of speech therapy may be questioned when communication skills are moderately to severely impacted in later phases of HD, our findings highlight the important associations between what was perceived as meaningful activities from the patient's perspective, and its possible positive impact on psychological well-being and quality of life (Schwartz *et al.* 2019).

Professional caregivers, and SLTs in particular, are also essential in ensuring that assessments of communication needs are conducted, in order to evaluate and determine whether and how patients with HD are able to communicate effectively. In the present study, few professional caregivers had received formal training in how to optimize communication by SLTs, regardless of repeated challenging experiences in communicating with patients.

AAC aids

AACs should ideally supplement the patient's ability to communicate, enable patients to feel they have an influence on their own situation and treatment, and facilitate the expression of thoughts, anxieties and reflections (Handberg and Voss 2018, Neto *et al.* 2017). The role and usefulness of AACs has received increasing attention during the past decade (Beukelman *et al.* 2007, Handberg and Voss 2018). Still, in our study, AACs did not seem implemented in the care of individuals with HD. This may be explained by insufficient knowledge about AACs in HD care settings, thereby reducing chances of use, in addition to a lack of training of relevant people surrounding the patient, professional caregivers, family and friends (van Walsem *et al.* 2016).

The need for communication aids will change during the progression of HD. Verbal and non-verbal communication skills will gradually decline and be lost, and in the later phases of the disease, AACs may be the only way for patients to express themselves (Ferm *et al.* 2010). People affected by HD should be encouraged to use speech and language supported by alternative communication as long as possible (Hamilton *et al.* 2012). Communication aids need to be implemented at an early stage of the disease, while patients are motivated, and before cognitive dysfunction has reduced the person's learning capacity (Hamilton *et al.* 2012). However, clinical experience and research (Hamilton *et al.* 2012) has demonstrated that patients in early phases of HD tend to consider themselves healthy enough not to need communication aids, as was confirmed in the present study. Professional caregivers also described the patients' lack of motivation as part of the problem. As an example, one of our participants with HD explicitly mentioned having no need for speech therapy. This participant's communication skills were, however, clearly impacted by HD (word searching, simple language, some problems with understanding), illustrating a possible lack of awareness of symptoms frequently observed in individuals with HD. This lack of awareness may pose a challenge for the use of communication aids, in contrast to other patient groups showing high acceptance rates to AACs, such as in amyotrophic lateral

sclerosis (ALS) or traumatic brain injury (Beukelman *et al.* 2007). Therefore, research on the appropriate timing of referral for AAC assessments and interventions, and how AACs could promote greater social inclusion and participation, remains an important challenge in HD (Beukelman *et al.* 2007).

Resistance to use AACs could also indicate a need to postpone considering any use of communication support in order to emotionally protect themselves from what they know is coming. Hence, help with processing the emotional burden of being affected by HD may be important in the earlier phases of the disease, to help them see the worth of learning to use a tool that could be useful for them later on. When introducing information about AACs to an individual with HD, considering how and when this should be suggested is crucial, and how the person best can be helped to accept an increased need for support (Patourel 1987, Scherer 2005, Scherer and Federici, 2015). The patient's attitudes regarding the potential use of AACs is important if its implementation is going to be successful, and needs to be followed up by the development of good strategies of how its use may fit into the patient's everyday life. Few studies have investigated this issue. Hence, future research is needed in this area.

There is a variety of existing AACs, ranging from low to higher technological levels. On the lower levels, we find communication books, with symbols or pictures supporting communication, and talking mats, to which pictures can be attached and rearranged as required, so that information can be presented in small chunks supported by symbols (Hamilton *et al.* 2012). Higher level AACs include software programs on computers or tablets that are specialized for reduced speech and language function, and may include photo-communication with speakers and audio functions. In order to find the right communication aid for a patient, the patient's communication needs, desires, abilities, cognitive strengths and weaknesses, in addition to environmental factors that could facilitate or complicate the use of AACs should be evaluated (Scherer 2005). A combination of different communication aids could also be necessary to cover the patient's different needs (Klasner and Yorkston 2001). Regular reconsidering of the communication aid is important, along with changes in the disease and new needs, in order to find the communication aid that the patient will be most comfortable using (Hamilton *et al.* 2012, van Walsem *et al.* 2016, Scherer 2005).

Clinical implications

In summary, several clinical implications follow from the present study. Professional caregivers working with

individuals affected by HD should receive information or guidance from a SLT in how to approach patients' difficulties with communication. Aspects of communication should also be a central and natural issue during multidisciplinary meetings. Implementing AACs in the care of individuals with HD requires that resources and time are allocated for the guidance and training of patients, relatives and staff. The need for AACs should be assessed at early stages of the disease, and close communication partners (family, friends and professional caregivers) should be included early in assessments, and in the implementation of interventions (Johansson *et al.* 2020).

In order to reach such aims, SLTs should be included in interdisciplinary teams, in line with the guidelines from the EHDN (Hamilton *et al.* 2012). Guidance and training by SLTs should be implemented in institutions' procedures and training of staff. Employing SLTs in care homes for patients with HD would strengthen the multidisciplinary team, but would also be important for patients, since it would secure that issues surrounding communication were integrated in the treatment and follow-up of the patients, potentially strengthening affected individuals' psychological well-being and quality of life.

Strengths and limitations

In this study, patients described challenges they experienced in the early and middle phases of the disease, whereas professional caregivers were involved in the care of patients in middle to later phases of the disease. Hence, the two groups' points of reference differed slightly. Nevertheless, we believe that results still complement each other. The number of participants in each group could also be seen as a limitation, especially given the high heterogeneity within each group. On the other hand, the aim of qualitative research being the investigation of in-depth subjective experiences, the current sample size was considered adequate. Dysfunctional speech and language skills in the patient group somewhat restricted the depth of the interviews, and data showed that some struggled with voicing their experiences, or gave conflicting responses. Nevertheless, we believe that including this group is a strength, giving a voice to people with first-hand experience with communication difficulties. An available, systematic and precise evaluation of the participants' speech and language skills would potentially have been optimal, but is rarely conducted in HD care settings. However, we do not believe that this information was central for the interpretation of results. Relatives and family were not included in the current study. Their inclusion would have broadened our knowledge about this patient group, and should be included in future studies. Finally, in-depth oral

interviews with the SLTs who provided written data would also have strengthened the study, but were unfortunately not possible.

Conclusion

The present study sheds light on patients' and professional caregivers' experiences of challenges with speech and language in HD, and suggests ways to improve communication in this group of patients. Communication with people affected by HD is challenging, but not impossible. Professional caregivers had developed strategies to enhance communication, but still expressed a need for more knowledge about how to facilitate conversations in patients under their care. The inclusion of SLTs into multidisciplinary teams responsible for the care of individuals affected by HD would serve both the patients' and professional caregivers' needs. Early implementation of AACs would have the potential to increase the patients' participation in daily life, and ease professional caregivers' care task. However, in order to achieve this, AACs need to be implemented while the patient is still motivated and has the cognitive capacity to learn, and should involve training of professional caregivers as well as relatives. More research is needed if we are to disentangle the complex challenge of optimizing and facilitating communication between patients with HD, their relatives, and professional caregivers.

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Data availability statement

Oslo University Hospital does not permit data to be publicly available due to privacy or ethical restrictions.

References

- BAIG, S. S., STRONG, M. and QUARELL, O. W., 2016, The global prevalence of Huntington's disease: a systematic review and discussion. *Neurodegenerative Disease Management*, **6**, 331–343.
- BAXTER, S., ENDERBY, P., EVANS, P. and JUDGE, S., 2012, Barriers and facilitators to the use of high-technology augmentative and alternative communication devices: a systematic review and qualitative synthesis. *International Journal of Language & Communication Disorders*, **47**, 15–29.
- BEUKELMAN, D. R. and BALL, L. J., 2002, Improving AAC use for persons with acquired neurogenic disorders: understanding human and engineering factors. *Assistive Technology*, **14**, 33–44.
- BEUKELMAN, D. R., FAGER, S., BALL, L. and DIETZ, A., 2007, AAC for adults with acquired neurological conditions: a review. *Augmentative and Alternative Communication*, **23**, 230–242.
- BILNEY, B., MORRIS, M. E. and PERRY, A., 2003, Effectiveness of physiotherapy, occupational therapy, and speech pathology for people with Huntington's disease: a systematic review. *Neurorehabilitation and Neural Repair*, **17**, 2–24.
- DALE, M. and VAN DUJIN, E., 2015, Anxiety in Huntington's disease. *Journal of Neuropsychiatry and Clinical Neuroscience*, **27**, 262–271.
- DOMARADZKI, J., 2015, The impact of Huntington disease on family carers: a literature overview. *Psychiatria Polska*, **49**, 931–944.
- FERM, U., SAHLIN, A., SUNDIN, L. and HARTELIUS, L., 2010, Using Talking Mats to support communication in persons with Huntington's disease. *International Journal of Language & Communication Disorders*, **45**, 523–536.
- GAGNON, M., BARETTE, J. and MACOIR, J., 2018, Language disorders in Huntington disease: a systematic literature review. *Cognitive Behavioral Neurology*, **31**, 179–192.
- GALTS, C. P. C., BETTIO, L. E. B. and JEWETT, D. C., et al., 2019, Depression in neurodegenerative diseases: common mechanisms and current treatment options. *Neuroscience Biobehavioral Reviews*, **102**, 56–84.
- GODEFROY, O., 2013, *The behavioral and cognitive neurology of stroke*, (Cambridge University Press).
- GRANEHEIM, U. H., LINDGREN, B. M. and LUNDMAN, B., 2017, Methodological challenges in qualitative content analysis: a discussion paper. *Nurse Education Today*, **56**, 29–34.
- GRANEHEIM, U. H. and LUNDMAN, B., 2004, Qualitative content analysis in nursing research: concepts, procedures and measures to achieve trustworthiness. *Nurse Education Today*, **24**, 105–112.
- HAMILTON, A., FERM, U., HEEMSKERK, A.-W., TWISTON-DAVIES, R., MATHESON, K. Y., SIMPSON, S. A. and RAE, D., 2012, Management of speech, language and communication difficulties in Huntington's disease. *Neurodegenerative Disease Management*, **2**, 67–77.
- HANDBERG, C. and VOSS, A. K., 2018, Implementing augmentative and alternative communication in critical care settings: perspectives of healthcare professionals. *Journal of Clinical Nursing*, **27**, 102–114.
- HARTELIUS, L., JONSSON, M., RICKEBERG, A. and LAAKSO, A., 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*, **45**, 381–393.
- HSIEH, H. F. and SHANNON, S. E., 2005, Three approaches to qualitative content analysis. *Qualitative Health Research*, **15**, 1277–1288.
- JOHANSSON, I. L., SAMUELSSON, C. and MÜLLER, N., 2020, Patients' and communication partners' experiences of communicative changes in Parkinson's disease. *Disability and Rehabilitation*, **42**, 1835–1843.
- JOHNSON, A. C. and PAULSEN, J. S., 2014, *Understanding Behavior in Huntington's Disease. A Guide for Professionals*, (Huntington's Disease Society of America).
- KLASNER, E. R. and YORKSTON, K. M., 2001, Linguistic and cognitive supplementation strategies as augmentative and alternative communication techniques in Huntington's disease: case report. *Augmentative and Alternative Communication*, **17**, 154–160.
- MCCOLGAN, P. and TABRIZI, S. J., 2018, Huntington's disease: a clinical review. *European Journal of Neurology*, **25**, 24–34.
- NETO, L. L., CONSTANTINI, A. C. and CHUN, R. Y. S., 2017, Communication vulnerable in patients with Amyotrophic Lateral Sclerosis: a systematic review. *NeuroRehabilitation*, **40**, 561–568.

- NEUMANN, S., QUINTING, J., ROSENKRANZ, A., DE BEER, C., JONAS, K. and STENNEKEN, P., 2019, Quality of life in adults with neurogenic speech–language–communication difficulties: a systematic review of existing measures. *Journal of Communication Disorders*, **79**, 24–45.
- NOVAK, M. and TABRIZI, S. J., 2011, Huntington's disease: clinical presentation and treatment. *International Review of Neurobiology*, **98**, 297–323.
- PATOUREL, J. L., 1987, *Communication aids for the progressively ill*. I P. Enderby (Red.), *Assistive Communication Aids—for the speech impaired*, 79–86. (London: Churchill Livingstone).
- POLLARD, J., 2008, *Hurry Up and Wait: A cognitive care companion: Huntington's disease in the middle and more advanced years*, (Lulu).com.
- POWER, E., ANDERSON, A. and 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*, **44**, 261–275.
- ROOS, R. A., 2010, Huntington's disease: a clinical review. *Orphanet Journal of Rare Diseases*, **5**, 40.
- RÜB, U., SEIDEL, K., HEINSEN, H., VONSATTEL, J. P., DEN DUNNEN, W. F. and KORF, H. W., 2016, Huntington's disease (HD): the neuropathology of a multisystem neurodegenerative disorder of the human brain. *Brain Pathology*, **26**, 726–740.
- SALDERT, C., FORS, A., STRÖBERGS, S. and HARTELIUS, L., 2010, Comprehension of complex discourse in different stages of Huntington's disease. *International Journal of Language and Communication Disorder*, **45**, 656–669.
- SCHERER, M. J., 2005, *Living in the state of stuck: how assistive technology impacts the lives of people with disabilities*. (Brookline Books).
- SCHERER, M. J. and FEDERICI, S. K. R., 2015, Why people use and don't use technologies: introduction to the special issue on assistive technologies for cognition/cognitive support technologies. *NeuroRehabilitation*, **37**, 315–319.
- SCHWARTZ, A. E., VAN WALSEM, M. R., BREAN, A. and FRICH, J. C., 2019, Therapeutic use of music, dance, and rhythmic auditory cueing for patients with Huntington's disease: a systematic review. *Journal of Huntington's Disease*, **8**, 393–420.
- VAN WALSEM, M. R., HOWE, E. I., FRICH, J. C. and ANDELIC, N., 2016, Assistive technology for cognition and health-related quality of life in Huntington's disease. *Journal of Huntington's Disease*, **5**, 261–270.
- ZAROTTI, N., SIMPSON, J. and FLETCHER, I., 2019, I have a feeling I can't speak to anybody": a thematic analysis of communication perspectives in people with Huntington's disease. *Chronic Illness*, **15**, 61–73.

Appendix

Interview guide for patients about Huntington's disease and communication

We want to hear about how you experience in communicating with other people. We want to hear about situations you experience as good and situations you may find challenging. There are no wrong answers.

The experience of communicating with other people

1. What does communication mean to you?

2. Do you experience differences in talking to people today compared to before you were diagnosed with HD?
- If yes: Could you say something about what kind of changes you notice when talking to your family?
 - When talking to friends and neighbours?
 - When talking to new people?
 - When talking to professional caregivers?
3. If you answered yes in question 2: How do you experience these changes?
4. When you talk to other people, are some situations more challenging than others?
- If yes: What kind of situations do you find most challenging?
5. Do you experience that you manage to express yourself so that other people understand you?
6. Do you experience that you understand other people when they are talking to you?
- If no:
 - a. Can you please give some examples of what would be difficult to understand?
 - b. Are there any situations you experience as more difficult when it comes to understanding what other people say?
 - c. What do you think and feel if you do not understand what other people say?
7. Is it easier to communicate with a person who has knowledge about Huntington's disease compared to a person who does not have this knowledge?
- If yes:
 - a. How do you experience the difference?
 - b. Who is the most difficult to communicate with?
8. What is important to you for communication to be as good as possible?

Experience with speech therapy and use of communication aids

9. Have you previously been in touch with a speech and language therapist?
10. Do you currently receive any follow-up by a speech and language therapist?
- If yes:
 - a. How often?
 - b. What kind of follow-up do you receive from a speech and language therapist? (For example: Adjusted food, communication?)
 - If no:
 - a. Is this your choice, or have you not been offered a follow-up from a speech and language therapist?
- If the patient has or previously had follow-up by a SLT (Yes to Question #10), ask question number 11. If no to Question #10, continue with Question #12.

11. Have you received follow-up by a speech and language therapist regarding communication?
 - If yes:
 - a. Was it useful? Why/why not?
 - b. What sort of follow-up did you receive? (For example guidance, ‘treatment’)
 - If no:
 - a. Would you have liked to be offered follow-up on communication?

Clarification of terms: A communication aid is an aid that compensates or supports reduction or loss of speech and language skills. Communication aids are divided into two overarching categories: High and low technology aids. High technology aids are for instance tablets with Cognitass. Example of low technology aids are for instance to use pen and paper or pictures when having a conversation.

- 12. Have you received any information about communication aids from a speech and language therapist or other professional caregivers?
 - If yes: Who has given you this information?
- 13. Do you use any kind of communication aids?
 - If yes: What kind of communication aid do you use?
 - If no:
 - a. What is the reason for not having tried communication aids?
 - b. Would you like to try a communication aid?
- 14. (If you have used or are using communication aids) Have you received training or received any follow-up in order to be able to use it?
 - If yes:
 - a. Who has given you the follow-up?
 - b. In what way have you received follow-up?
 - If no: Would you need any follow-up to be able to use the communication aid?
- 15. What benefit did you have/do you have from the communication aid?
- 16. Finally, do you have any thoughts about how speech therapists or other professional caregivers could improve communicate with you?

Thank you for taking the time to answer these questions!

Interview guide about communication for professional caregivers working with patients with Huntington’s disease

Information was collected about gender, age, workplace, profession and experience of working for patients with Huntington’s disease.

Communication experiences with patients affected by Huntington’s disease

1. What does communication mean to you?

2. During team meetings discussing patients, is the theme ‘communication’ on the agenda?
3. In which phase of HD is/are the patient(s) you are working with? (Early/middle/late phase)
4. How do you experience communication between you and the patient(s)? If you work with patients in different disease phases – do you experience variation across disease phases?
5. If you have known the patient(s) for a while, have you experienced any changes in the patients’ ability to communicate?
6. Do you feel/experience that conversations with the patient(s) are most related to physical/psychological needs, needs related to the disease, or to everyday needs?
7. Do you experience that you understand what the patient is communicating?
 - If you do not understand, what do you think is the reason for that? (For instance: Indistinct speech, volume, searching for words, mimicry, other).
8. Do you experience that the patient understands what you are communicating?
 - If yes: What could be the reason? (For SLTs: How do you know that the patient understands what you are communicating?)
 - If no: How do you know that the patient does not understand what is being said?
9. Is communication with the patient influenced by the physical setting? (For instance, the patients’ room, living room etc). (For SLTs: Please elaborate your answer instead of the follow-up questions beneath).
 - If yes:
 - a. In which way?
 - b. Are some communication settings better than others?
 - If no:
 - a. Do you always talk with the patient(s) in the same physical setting? (For example in the physiotherapy room)
10. What is important to you in order to optimise communication with the patient?

Experience with speech therapy and the use of communication aids

11. Have you collaborated or received guidance from a speech and language therapist regarding communication with patients with Huntington’s disease?
 - If yes:
 - a. Was it useful? Why/why not?
 - b. Which type of guidance did you receive? (Guiding, exercises, other?)
 - If no:
 - a. Have you collaborated with a speech therapist about for instance dysphagia/difficulty with swallowing with patients with Huntington’s disease?

b. Would you have wanted to get follow-up regarding communication?

(For SLTs: What kind of exercises do you perform with the patients with Huntington's disease? Which areas of difficulty are the exercises aimed at?)

(For SLTs: 12. Do you collaborate or have you collaborated with professionals and/or other speech therapists regarding communication with patients with Huntington's disease?

Please elaborate on your answer.)

Clarification of terms: A communication aid is an aid that compensates or supports reduction or loss of speech and language skills. Communication aids are divided into two overarching categories: High and low technology aids. High technology aids are for instance tablets with Cognitass. Example of low technology aids are for instance to use pen and paper or pictures when having a conversation.

12. Have you met patients with Huntington's disease who use or have used communication aids?

– If yes, approximately how many patients/how common is it?

13. If the patient uses a communication aid, have you experienced that it has been useful during your conversations with the patient? (For SLTs: Please elaborate why or why not it has been useful).

– If yes, in what ways?

– If no, in what ways and how do you explain this?

14. What kind of communication aids do the patient(s) use? Do you know why patients do not use communication aids?

15. Have you received any training in how to use communication aids with the patient? (For SLTs: Whom have you received trained from?)

16. Finally, do you have any thoughts about how professionals could improve communication with patients affected by Huntington's disease?

Thank you for taking the time to answer these questions!