Music therapy in Huntington's disease

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“La musique exprime ce qui ne peut être dit
et sur quoi il est impossible de rester silencieux”

Victor Hugo (1802 – 1885)

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Improving quality of life in patients with Huntington’s disease through music therapy

A qualitative explorative study using focus group discussions

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Abstract

Huntington's disease (HD) is a progressive, neurodegenerative, autosomal dominant disease, characterized by motor disturbances, behavioral and psychological symptoms and cognitive decline. These characteristics often result in loss of expressive and communicative skills, especially in the advanced stage of the disease, frequently giving rise to behavioral problems such as anxiety, irritability and apathy. Music therapy is potentially a good non-pharmacological intervention to improve communication skills and thus possibly reduce behavioral problems, leading to a better quality of life (QoL) overall.

As there is little knowledge on how music therapy may contribute to improve the QoL in HD, a qualitative research was conducted to gain insight: a focus group study was performed, following the guidelines of naturalistic inquiry and grounded theory. In three open-structured group discussions, participants (six HD professionals with different experience with and knowledge about music therapy) were asked to reflect on what defines QoL in HD and on the potential role of music therapy for clients with HD.

The results show that, according to the focus group participants, psychosocial aspects seem to be the most important aspects which contribute to improve QoL in HD-patients, with sense of security, confidence and structure being the qualifications most mentioned. Autonomy and self-esteem are other important QoL factors. The participants stated that the stage of the disease and the living conditions of the patient are crucial. Poor insight, unawareness and denial, all due to cognitive decline, mean that it is extremely difficult for patients to reflect on their own QoL. As the ability to communicate and express oneself deteriorates over time, music therapy could play an important role in the treatment of patients with HD in all phases of the disease. By providing an additional means of communication, thus enabling the patient to express emotions, music could be used as a mood-indicator to determine in which state of mind the patient is.

This article describes six HD-clinicians' perception of quality of life for patients with HD and the potential role music therapy can play in improving the QoL of patients with HD using focus group discussions. These insights were used to inform an empirical trial looking at the effects of music therapy on improving the quality of life of patients with HD.

Introduction

Huntington's disease

Huntington's disease (HD) is a rare autosomal dominant neurodegenerative disorder, caused by a mutation in the HTT-gene. This progressive disease is characterized by movement disturbances, neuropsychiatric disturbances and cognitive deterioration leading eventually to a complete dependency for daily life activities (Roos, 2010). Once manifest, the patient's life expectancy is about 15-20 years (Bates, Tabrizi, &Jones, 2014). Three clinical stages can be described: Stage I, in which patients develop initial symptoms but are still independent. Stage II, patients become more dependent as the disease progresses. Stage III is the end-of-life stage: patients become completely dependent for all daily life activities (Bates et al. 2014; Roos, 2010).

Many patients first present with neuropsychiatric or cognitive disturbances. Affective (mood) disorders (e.g. depression, anxiety, apathy, irritability) and behavioral problems (e.g. obsessive compulsive behavior and aggression) are often observed (Duijn van, Kingma, & Mast van der, 2007) and are recognized as being most distressing for both patients and their caregivers; they are often the main reason for institutionalization (Hamilton, Salmon, Corey-Bloom, Gamst, Paulsen, Jerkins, Jacobson, & Peavy,(2003) . Psychosocial stressors may include feelings of sadness and anxiety about the cognitive and physical decline and about changes in social role. They cause a great burden on the emotional well-being of patients with HD and their quality of life (A'Campo, 2012).

Verbal communication is often affected because of motor impairment of speech rather than a reduced desire for verbal communication (Ho, Robbins, Walters, Kaptoge, Sahakian, & Barker, 2004). A qualitative study on communication in HD, using interviews and focus groups with patients, family members and professional caregivers, found that all participants acknowledged the need for increased involvement in social life in order to enhance communication (Hartelius, Jonsson, Rickeberg, & Laakso, 2010). However, the gradual deterioration of communicative skills contributes to a decrease in functional health and an increasing inability to participate in various life situations. This may have a great impact on the quality of life of the patients, often leading to behavioral problems (Nance, Paulsen, Rosenblatt, &Wheelock, 2011).

Due to the progressive nature of the disease, cognitive losses accumulate throughout the different stages. The cognitive decline often results in behavioral problems and dysexecutive behavior (Ho & Hocaoglu, 2011). However, the patient's self-image doesn't seem to change throughout the stages. According to Ho, Robbins, & Barker (2006), patients with HD tend to underestimate consistently and persistently the degree of their dysexecutive behavior. There are three underlying components: cognition, self-regulation and insight. McCusker & Loy (2014) use the term "unawareness" when a patient's perception of obvious disease manifestations and impact differ from that of observers, such as clinicians or family members. Unawareness (or anosognosia), also described as denial and coping mechanism, correlates with disease progression, disease severity contributing to it to a more significant extent than disease duration (Sitek, Thompson, Craufurd, & Snowden, 2014).

To date, there is no cure for HD, nor can its progress be reversed or slowed down. All treatment must be individually tailored, as the symptoms and signs are different for each patient and change over time. The goal of all forms of treatment is to reduce any discomfort and pain, eventually leading to improvement of quality of life (Roos, 2010).

Quality of Life

One of many definitions of Quality of Life (QoL) is "...an individual's perception of his position in life in the context of the culture and value systems in which he lives and in relation to his goals, expectations, standards and concerns" (World Health Organization (WHO), 2002). QoL is multidimensional and includes domains that are related to physical, mental, emotional and social functioning and the social context in which people live (Ferrans, 2005). How these domains are related to patients with HD is illustrated in figure 1.

A distinction has to be made between the term QoL and Health-related Quality of life (HrQoL). The first term reflects the overall status of a combination of factors: a person's health, symptoms and level of physical and social functioning (Ready, Mathews, Leserman, &Paulsen, 2008). The latter is defined as "an individual's perception of his or her health and health-related domains of well-being" (WHO). In the present study we will use the term QoL indicating the health-related domains.

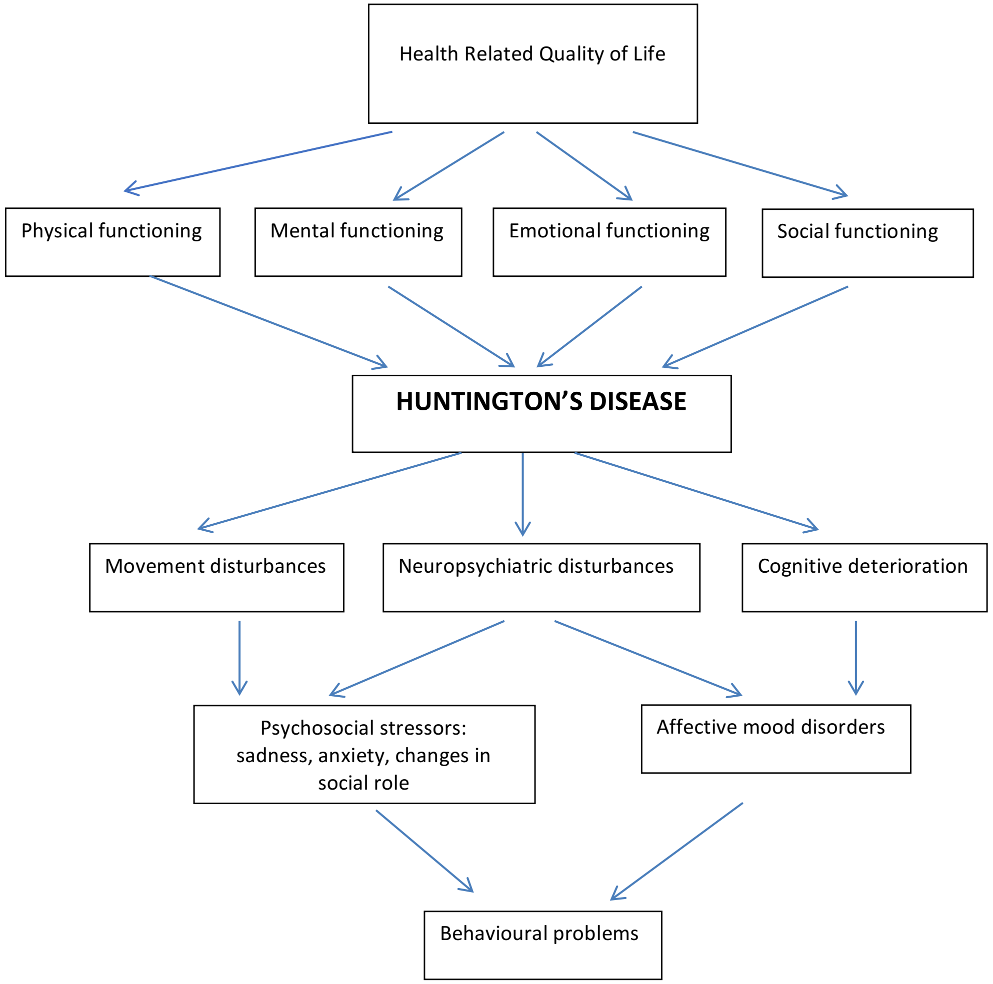

Figure 1: The four domains of health related quality of life in relation to Hungtington's disease

Quality of life and Huntington's disease

During the last few decades, several studies have been performed with the aim of determining which factors matter most with regard to the QoL of patients with HD. Helder, Kaptein, Kempen van, Houwelingen van, & Roos, (2001) concluded that physical and psychosocial symptoms have a great impact on the well-being of HD-patients, the latter being more severely affected. The accompanying motor, cognitive and personality changes can be held responsible for this outcome. It should be noted that in this trial the researchers restricted themselves to assessing motor and cognitive disabilities of HD-patients; no depression measure was included. Ho, Gilbert, Mason, Goodman, & Barker, (2009) conclude that depressive mood and functional ability are critically linked to QoL in HD. This outcome is underlined by Ready et al. (2008) who state that depression is a strong predictor of QoL in HD and that motor impairment, independent functioning and cognitive impairment had lesser yet significant associations with QoL. When focusing on the QoL and HD, there is reason to suspect that neuropsychiatric symptoms are particularly harmful. These symptoms are common in HD and include high rates of depression, suicide, obsessive compulsive disorder and personality change. Also, psychopathology tends to be more severe in HD (Ready et al., 2008).

In 2011, the European Huntington's Disease Network (EHDN) Quality of Life Working Group initiated a qualitative research on the impact of HD across the entire disease spectrum (Ho &Hocaoglu, 2011). Semi-structured face-to-face interviews were individually conducted with 31 persons living with different stages of HD, from pre-clinical gene carriers to advanced stage. Physical and functional decline peaked throughout the different stages, indicating that these issues were raised more frequently by the patients over the course of the disease. There were no significant changes between stages for the emotional and social domains. Cognitive impairments were only raised in the early and middle stages. In the advanced late stage, the cognitive impairments hardly featured any more. As mentioned previously, the patient's self-image is most probably the cause of this contradiction; it is the degree of cognitive impairment which affects insight at this stage. Also, in the late stages, the physical impairments became increasingly evident which makes determination of the cognitive, emotional and social domains extremely difficult (Ho &Hocaoglu, 2011).

Until a decade ago the QoL of patients with HD could only be assessed using generic QoL-measurement scales (Helder et al., 2001), (Hocaoglu, Gaffan and Ho, 2012). These generic tools are unlikely to capture the total and true impact of the more specific and unique consequences of HD, particularly the many non-physical aspects of the disease.

There has been little consistency in QoL models within the literature in the past 10 years. Providing an overview of the many concepts is beyond the scope of the present study. However, in the process of finding the concepts that best describe QoL in HD, the World Health Organization International Classification of Functioning, Disability and Health-model (WHO-ICF) is worth mentioning (WHO). This model is not specific to measuring HrQoL, it is more a mapping and classification framework than a guide for hypothesis generation in the area of HrQoL (Bakas, McLennon, Carpenter, Buelow, Otte, Hanna, Ellett, Hadler, & Welch, 2012). This model focuses on functioning (body-functioning and structures) and disability (activities and participation), but also addresses contextual factors (both environmental and personal). There are several disease-specific tools that measure QoL-related domains for patients with HD, such as the Behavioral Observation Scale Huntington (BOSH) (Timman, Claus, Slingerland, Schalk van der, Demeulenaere, Roos, & Tibben, 2005), the Problem Behaviour Assessment (PBA) (Kingma, Duijn van, Timman, Mast van der, & Roos, 2008), or the Unified Huntington's Disease Rating Scale (UHDRS) (Huntington Study Group, 1996). However, these tools all have limitations or lack sensitivity to marginal clinical effects in the later stages of the disease (Timman, 2005). The UHDRS-FAP (For Advanced Patients), developed in 2013 (Youssov, Dolbeau, Maison, Boissé, Cleret de Langavant, Roos, & Bachoud-Lévi) is more sensitive to change in the advanced stages of the disease.

A disease-specific patient-reported QoL outcome measure was developed in 2012 by Hocaoglu et al., 2012: the Huntington's Disease health-related Quality of Life questionnaire (HDQoL). The HDQoL consists of three sets of scales: (1) the primary scale representing the core factors underlying health-related quality of life in HD, (2) the specific scales representing clinically meaningful HD-specific profiles of pertinent aspects of health-related quality of life, and (3) the summary scale. The different scales that can be derived from the HDQoL can suit different clinical or research purposes. The HDQoL aims to fully capture the impact of HD on the everyday life of the sufferers. The HDQoL is valuable in promoting more fully informed decision-making in the management of HD, eventually leading to more effective treatments (Hocaoglu et al., 2012).

Music therapy, Huntington's disease and other neurodegenerative diseases

The literature on the use of music therapy and its efficacy in patients with Huntington's disease is limited. In a detailed literature review, Bruggen van-Rufi & Roos (2015) describe that music therapy interventions used to improve QoL for people with HD vary greatly. Most articles that were analyzed were derived from observational studies, were often small-scaled and the number of music therapy sessions was limited.

In none of the reviewed studies, was QoL the primary outcome. Music therapy interventions vary throughout the different stages of the disease. It is not the stage of the disease but the individual treatment goal which seems to determine the indication for music therapy. Precise aims and methods in relation to the stage of the disease have not been well established.

Since there is a paucity of literature about the effects of music therapy in HD-patients,
it is justified to search for such effects in other, similar neurodegenerative diseases, especially

dementia in general, dementia of the Alzheimer's disease (DAT) and Parkinson's disease (PD). Since the prevalence of these diseases in today's society is much larger than HD, there has been more extensive music therapy research into DAT and PD. However, comparison with these disorders needs to be done with some caution as these conditions have different impacts on the brain and central nervous system.

HD has some similarities with dementia and PD in that these disorders cause the brain, as in HD, to deteriorate progressively, affecting mental, cognitive and motor functions. Dementia (loss of mental abilities) is common in neurodegenerative disorders (Liou, 2010). Dementias are often classified by the region of the brain that is affected. PD and HD are sub-cortical based and AD is cortical based. Although this classification is rather controversial among researchers and physicians, some differences do exist, but there is disagreement on the degree of differences. Cortical dementia (AD) has more impact on semantic and episodic memory while patients with sub-cortical dementia (HD and PD) do not experience degradation of semantic memory per se; they find it challenging to accomplish cognitive tasks that require retrieval, but this will improve under the right conditions (i.e. being "cued") . Furthermore, sub-cortical dementias almost always result in motor disorders. In terms of cognitive effects, differences between dementias are still being studied (Liou, 2010).

The literature provides us with useful conclusions from scientific research investigating the benefits of music therapy with dementia and PD that may also apply for HD.

Music therapy provides stimulation and may help to prevent social isolation for elderly individuals suffering from more advanced dementia. If words are no longer recognized, familiar music may provide a sense of safety and well-being, which in turn may decrease anxiety (Brotons, 2000). Listening to music is significantly more effective in increasing levels of engagement and well-being than not being exposed to music, regardless of the level of cognitive impairment in people with slight to severe dementia (Sheratt & Thornton, 2004). In an in 2011 published updated Cochrane-review, an overall beneficial effect of music therapy with dementia could not be supported, but positive effects were found specifically in decreasing problem behavior (Vink, Bruinsma, & Scholten, 2011). In the recently update Cochrane review on music-based therapeutic interventions for people with dementia the authors conclude that music therapy may reduce depressive symptoms (Steen van der, Soest van-Poortervliet, Wouden van der, JBruinsma, Scholten, & Vink, 2017). Music therapy can decrease agitation and has a positive effect on enhancing communication and emotional well-being (Vink, Zuidersma, Boersma, Jonge de, Zuidema, & Slaets, 2012). Music therapy enables the recall of life experiences and

the experience of pleasant emotions. Through music, contact can be established, especially as language deteriorates during the later stage of the dementing process (Vink, 2013).

In a "state-of-the-art"-article on music therapy and Parkinson's Disease, Raglio (2015) concludes that interventions that involve music can offer important starting points in PD rehabilitation, effectively acting on motor, as well as non-motor symptoms. Although only a limited number of studies take the psychological outcome into account, an earlier research by Pacchetti, Mancini, Agilieri, Fundaro, Martignoni & Nappi (2000) showed a significant overall effect on motor improvement and beneficial effects on emotional functions, activities in daily living and quality of life. They concluded that MT is effective on motor, affective and behavioral functions.

In general, throughout the literature on music therapy and neurological disorders it is suggested that researchers need to focus on the remaining abilities of patients rather than focusing on the abilities that are diminishing (Van Beek, 2012). In a trial where the cognitive burden in HD is explored throughout the different stages, Papoutsi, Labuschagne, Tabrizi, & Stout (2014) support this idea by stating that "...enhancing cognitive reserve through cognitive interventions is very promising, because these interventions are noninvasive and readily available to be used (...) together with other more invasive therapeutic approaches for later disease stages. Such research has been limited thus far in HD (...) HD would be an ideal model to test such methods..." (Papoutsi, Labuschagne, Tabrizi, & Stout, 2014), p. 680).

Elaborating on this assumption, we decided to develop a focus group study to explore if music therapy could be such an intervention. The main purpose of the present study was to describe the perceptions of different HD-clinicians on music therapy. Understanding these perceptions will help us gain insight in the potential role the music therapist can play in the multidisciplinary team surrounding patients with HD.

Method

The focus group method is a qualitative research method to collect data using guided group discussions in order to generate a rich understanding of participants' experiences and beliefs. The discussions create a process of sharing and comparing among the participants, investigating issues of context and depth, and generate their own interpretations of the topics (Morgan, 1998). Powell & Single (1996) state that the focus group method is appropriate if

- a certain topic is very complex (as is QoL) and needs to be explored from different points of view.
- a large number of variables needs to be reduced to the most essential ones; since QoL is such a wide concept, the question what matters most in QoL was discussed extensively.
- little is known about a certain topic (music therapy to improve QoL); the focus group method has an explorative character and generates hypothesis.

Guidelines of two methods were followed: the naturalistic inquiry (Lincoln & Guba, 1985) and the grounded theory (Glaser & Strauss, 1967). The first is a method in which a series of techniques is used to conduct qualitative research. Using these techniques, the method emphasizes the trustworthiness of the trial, which is important in evaluating its value.

The second is a systematic methodology that is used to construct a theory through the analysis of data. On reviewing the collected data, recurring ideas and concepts become apparent. As more data are collected and reviewed, concepts become categories which form the basis for new theory.

The three main research questions were:
Based on your own clinical experience

- what is QoL in general for patients with HD?
- what are the most important aspects of QoL in patients with HD?
- Could music therapy play a role in order to improve QoL in patients with HD?

These questions were continuously repeated during all three meetings.

Participants

Six HD-professionals specialized in five different treatment areas participated in the study. They were recruited by way of homogeneous purposive sampling: they were chosen because of their rich clinical experience with HD. The sample consisted of one neurologist, one elderly care physician, one psychologist, one physical therapist and two music therapists. They were all working in The Netherlands but each in a different care facility specialized in HD-care, so none of them were direct colleagues. One participant had a working relationship with the principal researcher (the first author). The average age of the participants was 46 years and the average number of years of expertise with HD-patients was 15 years. Apart from the two music therapists, one participant had much clinical experience with music therapy, one had little experience and two had no experience at all. All six respondents had experience in scientific research: two of them had a PhD-Degree, two had a Master's Degree and two had a Bachelor's Degree.

Prior to the first meeting, the first author of this article visited all the participants separately and gave them extensive verbal and written information about the aim of the research, the methodology and the protocol. She handed out a topic guide in which she revealed the main topics of the discussions. The topic guide consisted of several articles about QoL (Helder et al., 2001, Ho et al., 2009 and Ho et al., 2011) and about music therapy (Bruggen van- Rufi, 2009 and 2010). She instructed all participants to read these articles before the first meeting, giving each participant the same level of knowledge to use as a starting point for the discussions.

Moderator and assistant moderator

The discussions were guided by a moderator. The moderator did not participate in the discussion itself but raised issues and encouraged participants to reflect on topics openly. She summarized what had been said during the discussion. The moderator was completely familiar with the questioning route and knew the rationale for each question (Krueger, 1997). An assistant moderator was also present to make sure all the equipment worked correctly and to make notes. After each meeting the moderator and the assistant evaluated the notes taken.

Procedure Data collection

We conducted a total of three focus group meetings lasting two hours, followed by a telephone call made by the moderator to each participant to enable them to reflect on the last (third) group meeting. The three research questions (see above) were the guideline throughout all three meetings and all topics were open to discussion.

The group-discussions were audio-recorded once all the participants had given informed consent. The recordings were transcribed verbatim by either the assistant moderator or the first author. Prior to each meeting, the participants were asked to write down their thoughts about the topics to be discussed. These papers were handed to the moderator and were accounted for in the analysis. Furthermore, notes were taken during each session by both the assistant moderator and the moderator. After the third meeting, no new themes seemed to emerge, leading to the conclusion that data saturation had been reached.

Trustworthiness

To ensure credibility and trustworthiness (two evaluation standards that are characteristic for the naturalistic inquiry approach), we used the following procedures:

Triangulation, member check, peer debriefing

We used different methods of data collection: focus group discussions and notes taken during the discussions (methodological triangulation). Also, as a form of data triangulation, prior to each meeting the participants were asked to write down their thoughts about the topics to be discussed, thus producing a written record of each participant's thoughts. Should time restrictions prevent them from expressing their thoughts during the meeting, these could still be accounted for in the analysis (authenticity). From the second meeting onwards, the moderator started by confirming that everybody had received the summary of the previous meeting and had been afforded the opportunity to give their comments, thus ensuring that no important information was missing and that they recognized themselves in the conclusions so far (confirmability, member check). During the analysis, the moderator examined the analysis process and records for accuracy (confirmability). All three audit reports were shared with all participants. These reports can be provided upon request. Furthermore, the preliminary results were presented and discussed with music therapists and neuroscientific researchers at an international music therapy congress (poster and oral presentation) (peer debriefing). Here, they challenged the interpretations of the data and the preliminary conclusions of the focus group study. Their objective feedback was used to finalize the conclusions of the study.

Analysis

The verbatim transcripts were analysed by the first author. The themes and concepts were derived from the data by using the open coding system: the descriptions used by the participants during the discussion were identified and codes that described the same themes were clustered or deducted. A content analysis was performed according to the constant comparison method, based on the grounded theory of Glaser & Strauss (1967). Data were extensively collected, coded and organized. In this approach the process of induction (determining what premises can logically be derived from the emerging themes) leads to deduction (what else would have to be true if these premises are true). The whole process of constant comparison leads to general conclusion(s) which can logically be derived from the data (Boeije, 2005), (Copi, Cohen & McMahon, 2015).

Throughout the whole analysis process the codes and (sub)categories were checked with the moderator. The participants received a summary of every meeting, describing the (preliminary) analyses and highlighting the key findings. They were asked to check the content (member check/member validation), in certain instances resulting in modifications (negotiate outcomes).

Results

The results presented below first describe the participants' perceptions of

- QoL in general for patients with HD the most important aspects of QoL in patients with HD
- Subsequently, elaborating on the results of the first two research questions, the participants' perceptions on the third research question will be presented:
- the potential role of music therapy in improving QoL in patients with HD.

In the discussion we will expand on these interpretations in more depth, reflecting the participants' perceptions, tying together their ideas with the rationale for music therapy with patients with HD from the literature.

Q: What is QoL, based on your own clinical experience?

Analysis of the verbatim transcripts revealed several themes and subcategories (see fig. 2) which are presented below, illustrated with the most distinct quotes, accompanied by a short interpretive summary.

In each of the three sessions, the participants cited a number of items. The one theme being mentioned most often was the subjectivity of QoL:

> *"...One patient can have a very low self-esteem because his involuntary movements are so conspicuous. Another patient doesn't mind that his motor functions are poor, as long as his cognitive skills are still good. No matter how badly a person moves, he can still feel good about himself... "*

> *"...the only person who can say something about it is the patient himself. We, the professionals, can only ask, or guess..."*

A typical comment where all participants agreed upon was that patients have difficulties reflecting about their own QoL due to the cognitive decline. Besides that, one of the participants felt that patients do not express themselves in QoL-terminology, because in his opinion, QoL is a term used by the professionals, who must listen beyond what has actually been said and "translate" it into QoL-issues. All participants agreed that this "translation" might not fully reflect the patient's perception of his own QoL, as it is the interpretation of the professional. Elaborating on this, another comment was that the professionals tend to be too protective towards the patients:

> *"…Life consists of ups and downs, that is part of life. We cannot and must not protect our patients from that…"*

All participants agreed that recognition of the individual needs of the patient is most important when thinking about his QoL. They all emphasized that functionality and autonomy of the patient himself must be maintained and supported as long as possible. Involving family members in achieving this was also recognized by all participants as being very desirable.

One of the participants who is involved with patients at the earlier stage of the disease, before admission to a nursing home, or even before onset of the symptoms, stated:

> **"***…Quality of life is not an issue that is raised frequently in the doctor's office because the doctors have only little to offer in that aspect. The emphasis should be on care, and not on cure…"*

Following this statement, the discussion continued more specifically in the direction of QoL and the disease itself.

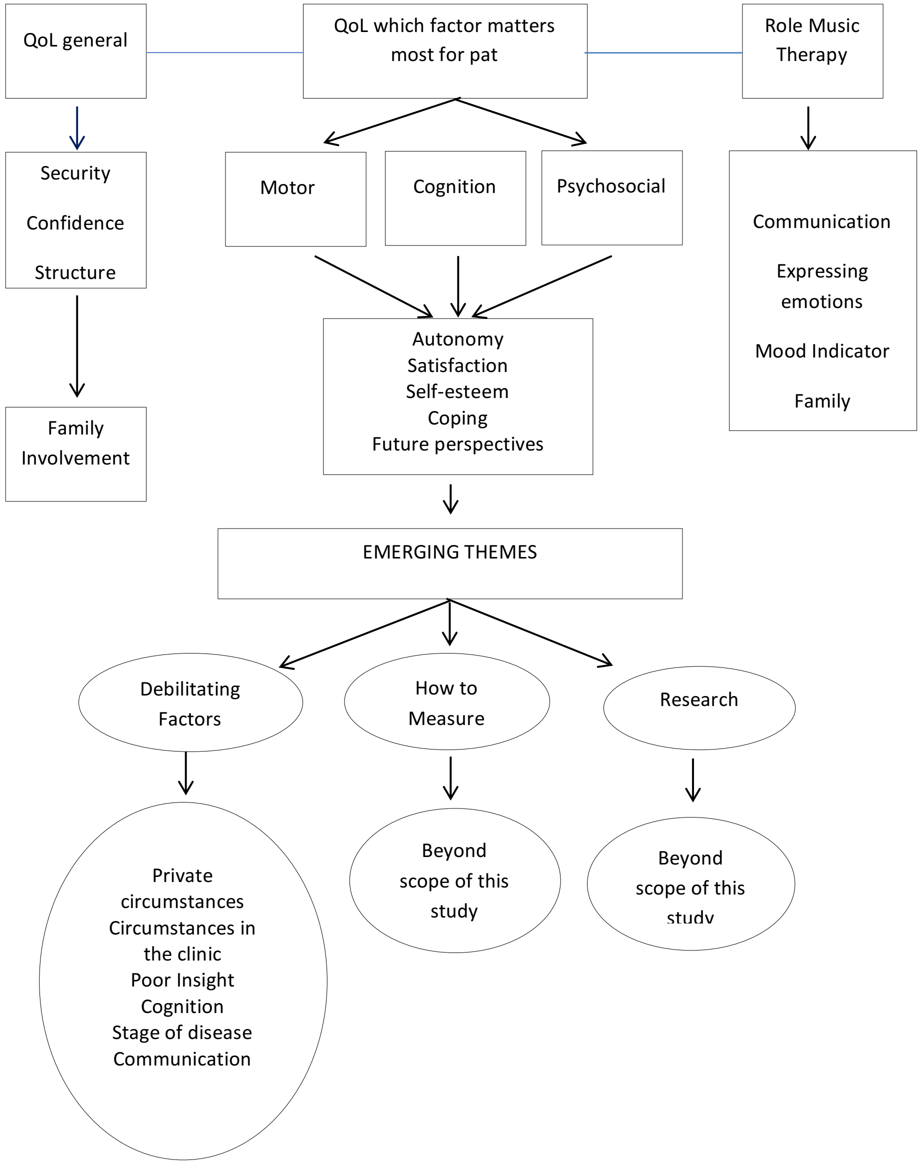

Figure 2: (sub)categories and emerging themes

Q: What are the most important aspects of QoL in patients with HD?

The content analysis of the transcripts revealed that, according to all participants, psychosocial and cognitive symptoms influence QoL most. Three major themes were identified: stage of the disease (1), cognitive decline (2) and circumstances/living conditions (3). While discussing these major themes, three subcategories emerged: (sense of) security (a), confidence (b) and structure (c).

In fig. 2 the findings are summarized in subcategories under each theme. How the subcategories relate to the three major themes is illustrated in fig. 3. In fig. 4, the consequences of the cognitive decline in relation to self-reflection about QoL is highlighted. Together they form an overall view of the participants' ideas of (health-related) quality of life and HD.

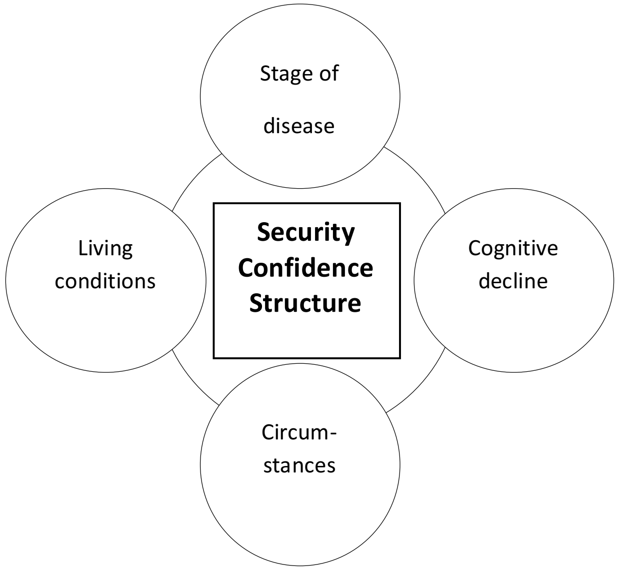

Figure 3: Relation subcategories and major themes

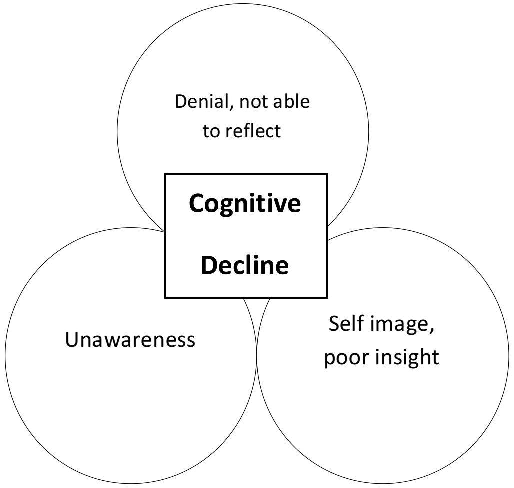

Figure 4: Consequences of cognitive decline i.r.t. self-reflection

Stage of the disease and cognitive decline

All participants agreed that the stage of the disease and the cognitive decline are inextricably related. The cognitive deterioration becomes more distinct as the disease progresses. All participants recognized the patient's inability to reflect about his own QoL. Denial, poor insight and poor coping skills were mentioned as the most debilitating factors in being able to do so adequately:

> *"...in the earlier stage of the disease, the patient tends to deny the symptoms and signs while in the later stage the patient can no longer reflect about it..."*

> *"...Denial comes from the patient himself, while poor insight is a result of cognitive decline in the later stage of the disease. Self-image does not grow with the progression of the disease, often resulting in behavioral problems..."*

One participant shared her experience with the group: when asking the patient about how he feels, he will often score himself "average" on a rating scale, adjusting to new circumstances.

> *"...Everybody has his own highs and lows, no different in patients with HD. The average will always be in the middle..."*

The discussion that followed, as one of the participants raised the question how HD-patients react differently from (healthy) persons in certain circumstances, focused specifically on the cognitive decline of patients with HD.

One of the participants commented that the cognitive decline can be beneficial for the patient:

> *"...it can be a relief for the patient as he is no longer aware of his decline. He is coping with and adjusting to his new situation..."*

Following this comment, the participants agreed that the cognitive decline results in yet another paradox: the impact of the disease seems to be less of a burden for the patient than for the people around him. This can cause much stress, especially when patients still live at home. All participants agreed that family support is very important. More specifically, they agreed that all people surrounding the HD-patients must have a comprehensive knowledge of the disease in order for the patient to feel confident and secure.

The discussion continued with these emerging themes on security, confidence and structure. One of the participants stated that

> *"...sense of security and confidence come from within the patient himself; structure must be provided by the people around the patient. The conditions and circumstances he lives in must be created by the people surrounding him..."*

This statement resulted in the unanimous recognition that another important distinction has to be made: Is the patient still living at home, does he receive daily care from family members, does he visit a day care facility or is he living in a long-term care facility.

Circumstances/living conditions
As the cognitive decline affects the patient's ability to structure his own life as the disease progresses, he becomes more and more dependent on his surroundings. One of the participants shared his perception with the group by commenting that

> *"...structure is the basis of feeling safe. We have to make sure that patterns, rituals, everything that the patient is used to, continue every day..."*

Following this comment, the discussion continued with reflecting about structure.
Participants agreed that structure seems to be best secured by living in a long-term care facility that is specialized in HD care because of the specific expertise these facilities have.

> *"...it takes very much time for a patient with HD to build trust. The more knowledge and affinity you have, the sooner this trust is a fact. This is an absolute necessity to treat the patient effectively..."*

However, once admitted to a LTCF, autonomy, self-esteem and future perspectives for the patient are QoL issues which are raised more often. The discussion continued while reflecting on these emerging themes:

> *"...each new situation results in severe tension and stress. Processing new information is extremely difficult, due to cognitive decline of the disease in the later stages..."*

> *"...the patient sitting in front of us is someone who still wants to be heard. A person with needs and wishes like all other human beings..."*

When autonomy is jeopardized, the participants agreed that living in a LTCF specialized in care for HD-patients is most desirable. They will find themselves surrounded by specialists, resulting in an improved feeling of security and confidence.

> *"...Even though the patient might not want to be here, he feels at ease, safe. All the people around him, fellow patients and nursing staff, know what he is going through..."*

When no more new emerging themes seemed to be raised by the participants, the monitor summarized the results so far of the group discussion. While all participants recognized that the subjective nature of the QoL-concept requires self-report by the patient for as long as possible, it was agreed that this becomes more and more challenged due to disease progression. When the HD-patient loses his ability to express himself in the later stage of the disease, other means of communication should be provided. The monitor used this conclusion to introduce the last and foremost important question, encouraging the participants once again to reflect open minded and freely:

Q: Can music therapy play a role in improving QoL in patients with HD?
Before going into the different themes, it is worth mentioning that some general yet distinctive "opening"-thoughts were shared by some of the participants on which the rest of the discussion elaborated:

> *"...as long as there is still no medication to cure the disease we should try every possible (non-pharmacological) intervention and treatment. Music therapy could very well be one of them (...), well worth giving it [MT] a try..."*

Elaborating on the aforementioned themes that emerged from the first two QoL-questions, three major themes were found: new ways of communication and expression, mood-indicator and family-participation *(*see fig. 5). Below, the participants' perceptions on these themes will be presented and quotes will be used to illustrate each theme.

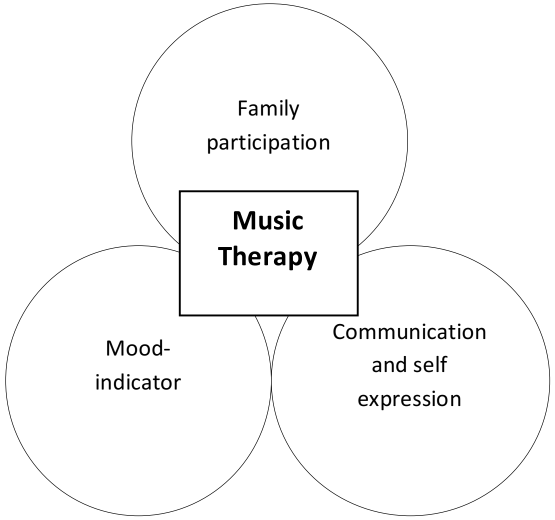

Figure 5: Potential roles of music therapy

Communication and expression

One of the participants felt that if a person loses the ability to communicate with others, music could take over:

> *"...music could be a new way to communicate again and to come to the inner self of the patient, to his emotions. The non-verbal character of music allows the patient to express himself in a different way (...) when a person can no longer give words to his feelings, music can give him new ways of expressing himself..."*

Another participant added:

> *"...Within music therapy the patient can express himself both non-verbally (playing musical instruments) and verbally (writing song lyrics, listening to existing songs and reflecting about the lyrics, "translating" them to his own life). In doing so, he gives meaning to his own emotions..."*

Mood indicator

Following these thoughts, the discussion continued when a new theme emerged: the use of music as a mood indicator. Some of the participants shared their own personal experience with music. Another participant who has no affinity with music whatsoever shared her thoughts as well, directing the discussion about the benefits of music in both positive and negative ways. However, putting their own personal thoughts about music aside, all participants agreed that for some patients music could be helpful.

One of the participants recognized the potential role of music, stating that

> *"...music could be used as an indicator to determine in which state of mind a patient is..."*

> *"...in music therapy the patient can create his own song, record it and give this to his family members. In this way the family could not only recognize the mood the patient is in, but also the person behind the patient, the partner or the father he once was, and still is or wants to be..."*

The participants agreed that even though the patient might no longer be able to reflect on the state of mind he is in, his choice of music or his reaction to the music played for him might help him express his feelings and help the people around him understand the patient better.

This last thought led the discussion to yet another emerging theme:

Involving family members

In relation to improving the QoL, all participants agreed that involving the direct family members or other caregivers is desirable.

"...the family relation between the patient and his partner and/or his children can be reinforced through music therapy..."

"...within music therapy partners could relive good experiences from before the disease kicked in hard (...) but also they can experience what is still there..."

"...music therapy could be a resource to give a positive change to an often problematic relationship. The healthy partner can recognize the "old" partner again..."

Discussion

This study is a first attempt to highlight the different opinions of a heterogeneous group of experienced professionals about quality of life in (advanced) Huntington´s disease, and the possible role music therapy could play. The main purpose of the research was to describe different clinicians' perceptions of music therapy in improving the quality of life in patients with Huntington's disease. Six professional clinicians from varying disciplines, all specialized in HD, discussed three major themes: QoL in general, QoL in HD and music therapy for HD-patients. In the discussion below we will first expand on the participants' perceptions about QoL, tying together their ideas with the literature. Subsequently we will do the same for the perceptions of the participants about music therapy.

In general, it was stated that reflecting about a patient's QoL, when he is incapable of doing so himself (as is the case in most HD-patients, especially in the later stages), is extremely difficult, given the subjective nature of the topic. Participants agreed that functionality and autonomy of the patient are the most distinct QoL-factors. When focusing on QoL of HD-patients, psychosocial and cognitive symptoms seem to influence QoL most. Stage of the disease, cognitive decline and the circumstances/living conditions of the patient are being recognized as the most important determinants of QoL.

Furthermore, HD-patients face many psychological issues such as anxiety, stress and hopelessness. Once their symptoms have become manifest, the patients realize their future perspectives, which has an immense impact on their QoL. As a result of the inability to express oneself due to the lack of (verbal) communication skills, it is highly likely that psychosocial problems will occur (Vervoort & Zuuren van, 2009).

The patient should feel secure, safe and confident. Sense of security and confidence are intrinsic factors and come from within the patient himself. However, structure in his daily life and the conditions and circumstances in which he lives seem to be decisive and are extrinsic factors which must be provided by the people surrounding the patient. In short, interaction between patient and caregiver is crucial, inevitably causing yet another area of stress (Vervoort & Zuuren van, 2009).

Determining which aspects are most important with regard to the (health-related) quality of life of patients with Huntington's disease is hindered by the many debilitating factors surrounding the patient, both physically and psychologically.

Because of this, it is justified and common to rely on the report of the caregivers and other proxies of the HD-patient, even though "...the subjective nature of the concept of QoL requires its assessment to be carried out by self-report where possible..." (Hocaoglu et al, 2012, p. 1793).

There is, however, the risk of being too paternalistic and overprotective. McCusker & Loy (2014) state that "...the primary ethical dilemma for all care providers is the balance between a patient's family's concern regarding care and safety and maintenance of the patient's autonomy, privacy and engagement..."(p. 6).

Relying on proxies presents another dilemma to be taken into account: many people with serious and persistent disabilities experience a good or excellent quality of life, while to external observers it seems these individuals live an undesirable daily existence. The impact of the disease appears to be less of a burden for the patient than for the people around him. This so-called disability paradox can also be recognized by people with HD (Albrecht & Devlieger, 1999).

In short, the different perceptions about QoL of the focus group participants are consistent with the literature. While there are many debilitating factors that influence the QoL in HD, it is important to find a concept that best describes all these disease-specific issues.

As detailed previously, the discussion about the potential role of music therapy in improving QoL for HD-patients resulted in three major themes: new ways of communication and expression, mood-indicator and family-participation (see fig. 5). These findings are consistent and can be confirmed with the music-therapy literature.

Communication and expression

Improving communication skills of individuals have been frequently reported in music therapy (McFerran, Lee, Steel, & Bialocerkoswki, 2009, Bruggen van – Rufi, 2009). As the ability to communicate and express oneself deteriorates over time, music therapy could play an important role in the treatment, in that it could provide an additional means of communication. The results from a music therapy referral audit conducted by Daveson (2007) show that patients with HD were most likely to be referred to music therapy due to a need for emotional expression, and the maintenance of communication skills and social relationships. This outcome can be confirmed by Bruggen van – Rufi (2010) who interviewed 14 members of the multidisciplinary team in a nursing home specialized in HD in The Netherlands. In this research, too, communication and self-expression were mentioned most often as treatment goals for referral to music therapy.

Mood indicator

Due to the affective (mood) disorder that is often manifested in the later stages of the disease, people suffering from HD have a diminished ability to recognize and express emotions, both in themselves and in others. Music and emotion seem to be inextricably connected (Juslin & Sloboda, 2010). Musical experiences can empower people to feel that they are alive and also promote shared experiences with others (McFerran et al., 2009).

Family participation

Music therapy can be offered both individually and in group sessions, with fellow-patients and with family members (Bruggen van-Rufi & Roos, 2015). As the focus group stated that involving family is of prime importance, the beneficial role of music therapy can be confirmed by McFerran et al, 2009; they conclude that using music therapy interventions to help improve communication skills with their carers outside of the music therapy sessions will be the most beneficial outcome.

In conclusion, because of the changing needs of the HD-patient, inherent to the stage of the disease, the treatment itself has to be adjusted to the individual needs important at that specific moment (Davis & Magee, 2001, Daveson, 2007). In HD the stage of the disease is crucial; while in the early stage denial might be an issue, in the middle and late stages poor insight into the disease itself might be the reason why QoL issues are raised less frequently by the patient. In the later stage of the disease, the patient might no longer be able to reflect about his own QoL.

Although the literature shows that precise aims and methods in relation to the stage of the disease are not well established yet (Bruggen van-Rufi & Roos, 2015), the focus group results suggest that music therapy may be beneficial throughout the various stages. Music therapy can combine functional training with cognitive, emotional and communicative/relational approaches. In so doing, the music therapist can forge a link between medical and psychological treatment. Methods and techniques can be constantly adapted, guided by the individual treatment goals of the patient, bearing in mind all of his or her limitations and skills. In short, the sessions can be “tailor-made” to the patient and his needs (Bruggen van-Rufi & Roos, 2015).

Strength and limitations

The strength of our study lies in the richness of experience of our participants. Collaboration with other professionals is imperative in order to meet the patient's needs. Except for the two music therapists, all participants came from different disciplines, and each was working in a different health care institution. Also, all of them had many years of experience with Huntington patients and had experience with or knowledge about scientific research. They represented the well-being of the patient with HD in every aspect. Furthermore, the number of participants was small enough to allow us to examine their responses in depth. They needed little encouragement to describe their experiences in detail.

At the same time however, having only six professionals participating in the study is a major limitation. Although the participants contributed a variety of different opinions, different issues might have been raised if more clinicians would have participated. Also, all participants were from The Netherlands. It is possible that clinicians from other countries may have different opinions, especially because long term care facilities (LTCF) where only HD-patients reside are not so common outside The Netherlands.

Another weakness of the research might have been the involvement of two music therapists with the four other clinicians who may have felt inhibited by their presence to provide negative views about music therapy. The reason for this was to make sure that at least one music therapist would be present at every session, anticipating possible absence of one of them. We cannot rule out the possibility that both music therapists were, unintentionally, advocating the use of music therapy during the group discussions, or stimulated socially desirable answers. We tried to minimize this by instructing them beforehand not to *advocate* but to *inform about* music therapy. This was a pragmatic choice**,** however we are well aware that instructing participants is contradictive for the naturalistic method. This is another weakness in the study.

The results obtained in this research must be regarded in the light of some more limitations. First of all, the participants were all acquainted with the main researcher and were aware of her ideas about setting up a music therapy research for HD-patients. For this reason, the researcher herself was never present at the meetings, nor talked about the trial while meeting the participants on different occasions.

Also there could be concern that the “must-read-articles” about music therapy, written by the main researcher, could have influenced the participants and led them to be more positive about music therapy and to conceal their criticism about music therapy.

Another weakness of the study was that in the first two meetings not all participants were present. However, the absent members were all given sufficient opportunity to participate in the group discussions by inviting their comments on the preliminary reports of each meeting, sent to them prior to the next session. At the start of the next meeting, those who had been absent from the previous meeting were given the opportunity to ventilate their thoughts. All participants were present at at least two of the three meetings.

Finally, in the attempt to find a representative and homogeneous sample where participants had a certain comparable level/degree of (scientific) experience, expertise and knowledge, we decided not to involve patients, caregivers or nurses. The results of this research reflect the opinions of professionals, not of the patients or caregivers themselves. Taken the aforementioned pitfalls into account (unawareness, disability-paradox, etc.), an additional focus group study in which patients and caregivers are the participants is recommended.

Although over the last few decades there has been an increasing number of clinical trials focused on the search for new therapeutic (pharmaceutical) strategies to manage HD, unfortunately few advances have been made. While there is still no cure, the emphasis should be on the care of patients with HD, especially in the advanced stage of the disease (Roos, 2010). Contributing to this need, a multi-center randomized controlled trial studying the effect of music therapy in Huntington’s disease to improve the quality of life of patients with HD, by way of improving communication skills, was conducted from October 2014 until June 2016 in four different long term care facilities in The Netherlands (Bruggen van – Rufi, Vink, Achterberg, & Roos, 2016). The results of the present focus group study helped informing this RCT. Final results of this study are expected in the winter of 2017.

Future prospects

Until recently, there was a lack of a disease specific measurement tool to assess music therapy with HD. Such a tool, the Music therapy Assessment Tool for Advanced Huntington’s Disease (MATA-HD), has now been developed. The results of a pilot study to validate this tool were recently published (O’Kelly & Bodak, 2016). Preliminary data indicate that the MATA-HD is a promising tool for measuring patient responses to music therapy interventions across psychological, physical, social, and communication domains of functioning in patients with advanced HD.

Conclusion

The results of the present focus group study suggest that music therapy can target many facets that are important to the QoL of patients with HD. As the ability to communicate and express oneself deteriorates over time, music therapy could play an important role in the treatment of patients with HD in all phases of the disease. Through music therapy an additional means of communication can be provided, enabling the patient to express his or her needs and emotions. Also, music can be used as a mood-indicator. Finally, involving family members in the treatment is desirable and should be encouraged. In music therapy this can be easily accomplished .

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