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**Title:** Wishes for the end of life in Huntington's Disease. Observations and reflections, initiated in The Netherlands  
**Issue Date:** 2014-11-12
Euthanasia and advance directives in Huntington’s disease: Qualitative analysis of interviews with patients

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Verena Rödig
Dick P Engberts
Aad Tibben
Raymund AC Roos

ABSTRACT

Background: In the literature there are few reports on euthanasia or physician-assisted suicide (PAS) or other matters concerning the end-of-life in patients with Huntington’s disease (HD), although clinical experience suggests these issues do arise.

Objective: To obtain in-depth information about patients’ thoughts on and attitudes to euthanasia, PAS and the use of advance directives in HD. To assess the difficulties patients encounter when thinking about end-of-life wishes.

Methods: Semi-structured in-depth interviews with 14 unselected HD patients from our out-patient clinic based on a topic list. Qualitative analysis of the interviews based on grounded theory.

Results: We identified three patterns in our group of respondents: patients with distinct wishes, with general wishes and ideas and patients with no wishes. The most important frame of reference regarding end-of-life wishes in HD patients or known gene carriers is the experience with an affected parent. Family is important when thinking about the end of life and advance directives, even more so than the patient’s physician. Knowledge about the (requirements of) law is limited.

Conclusions: The majority of interviewees expressed some kind of wish regarding end of life, probably more than they had revealed to their physician, but were sometimes hesitant to discuss it. Knowledge on how to deal with wishes, advance directives and response shift is limited. In general, patients underestimate the requirement for sound professional support when considering euthanasia or PAS and the value of an advance directive. In an attempt to improve knowledge and communication about end-of-life issues, physicians should ask the patient directly about their wishes.
INTRODUCTION

Huntington’s disease (HD) is an autosomal dominant progressive neurodegenerative disease, characterised by chorea and hypokinesia, psychiatric symptoms and progressive cognitive decline leading to dementia. To determine if a patient carries the HD-gene DNA-testing can be performed, either in the premanifest or manifest phase. There is no cure and all patients will eventually become totally dependent for all daily life activities. The primary cause of death is pneumonia, the second cause is suicide, the rate being 5-12 times higher in the HD population than in the general population. Prevalence in The Netherlands is approximately 1,700 patients; some 5,000-8,000 persons are at risk.

Euthanasia or physician assisted suicide (PAS) has been legal in The Netherlands, albeit under strict conditions, since the euthanasia act was approved by parliament in 2002. Approximately 7% of Dutch inhabitants have an advance directive, usually a euthanasia request. This document is not a patient’s right, nor is it part of normal medical treatment. It is a request, not a binding contract with legal consequences.

In 2011, approx. 2.5% (n=3,695) of all deaths in The Netherlands were the result of euthanasia or PAS; in approx. 85% the underlying disease was cancer. The reason for most people in the Netherlands creating an advance euthanasia directive is to make arrangements in case dementia develops. Symptoms and signs leading to a euthanasia request in Amyotrophic Lateral Sclerosis (ALS) were dependency, being limited in communication and anxiety, whereas in cancer patients the reasons were pain and fatigue. In 2005, about 8,400 requests for euthanasia were made, 2,400 of which were granted. Several situations arise for non-performance: the patient dies before euthanasia is granted, in 13% of the cases the request was withdrawn and 12% of the unperformed requests were refused by a physician. According to physicians the most debated and difficult requirement of the law on the basis of which the request should be judged is the requirement of unbearable suffering, which has subjective aspects.

In The Netherlands, between six and ten euthanasia requests from patients with HD were granted each year between 2007 and 2011. Besides anecdotal reports there are few studies on euthanasia or PAS or other end-of-life questions and the content of wishes for the end of life in patients with HD. HD differs from ALS and cancer with regard to disease duration and cognition. A third difference is that many HD patients and mutation carriers know the characteristics and the course of the disease from a family member and thus
have a precise idea of what their future will be like, usually at a much earlier stage than patients with other diseases.

With an increasing prevalence of neurodegenerative diseases and usually limited treatment options, the possibility of deciding the time of one's death by euthanasia or PAS could be a subject of interest for expanding treatment or guidance options. This study therefore aimed at gaining insight into the thoughts and attitudes of patients regarding end-of-life issues and the use of advance directives. We also examined which difficulties patients experience when thinking about end-of-life wishes.

METHODS

Participants

All patients who visited our out-patient clinic in the first half of April 2011 (n=19) were asked to participate in an interview about their end-of-life wishes; 14 consented. The reasons for declining were for four patients 1) “Life is perfect at the moment” or “I do not have any wishes because I take life as it comes”, 2) for one patient the reason was a recently developed depression.

Interview procedure and data analysis

Semi-structured interviews were conducted. We interviewed the patients in their home or in our out-patient clinic. The study was based on grounded theory. Initial interview topics were formulated after examination of the relevant literature. Consistent with standard qualitative research techniques, the interviews were based on a topic list, which evolved as the interviews progressed through an iterative process to ensure that the questions captured all relevant emerging themes. The interviews focused on thoughts and attitudes towards the end of life, the presence of advance directives, the experiences with HD in the family, and conversations about end-of-life wishes with either family members or a physician. Thematic saturation was reached after the 12th interview. All interviews were performed by the first author (SB) and took approximately one hour to complete. All interviews were recorded and transcribed verbatim. As our study was explorative, we used open coding. The transcripts were read and categorised into similar subject areas using inductive coding by SB and VR. Examples of codes are illness and
experiences in the past, importance of family and the role of a general practitioner, quality of life and threshold of acceptable life. The list of codes was discussed by SB and VR.

RESULTS

Characteristics of patients

The patients ranged in age from 28 to 70 years; eight participants were female. Five patients went to a day-care facility or lived in a nursing home. Two patients were in the premanifest phase, 12 patients in manifest phase, three in stage 1, three in stage 2, four in stage 3 and two in stage 4. Mean Total Functional Capacity (TFC) was 9.5^a (Table 3.1).

Background of end-of-life wishes

Most patients refer to their experience with the affected parent or other relatives and speak about it frequently and spontaneously. For many patients this has resulted in an intention to exert control over the end stage of the disease and the reason for having

<table>
<thead>
<tr>
<th>Patient/ respondent</th>
<th>Gender</th>
<th>Age</th>
<th>Marital status</th>
<th>Advance directive</th>
<th>Wishes</th>
<th>TFC</th>
<th>Disease stage</th>
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<td>No</td>
<td>Yes</td>
<td>13</td>
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</tr>
<tr>
<td>3</td>
<td>Female</td>
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<td>Relationship</td>
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<td>Yes</td>
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</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>43</td>
<td>Married</td>
<td>Filling in papers</td>
<td>Yes</td>
<td>11</td>
<td>1</td>
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<tr>
<td>5</td>
<td>Male</td>
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<td>Married</td>
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<td>Yes</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>6</td>
<td>Male</td>
<td>67</td>
<td>Divorced, ex-wife care giver</td>
<td>Yes</td>
<td>Yes</td>
<td>3</td>
<td>3</td>
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<tr>
<td>7</td>
<td>Female</td>
<td>68</td>
<td>Married</td>
<td>No</td>
<td>No</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>8</td>
<td>Male</td>
<td>44</td>
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<td>No</td>
<td>Yes</td>
<td>11</td>
<td>1</td>
</tr>
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<td>9</td>
<td>Female</td>
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<td>3</td>
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<td>No</td>
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<td>66</td>
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<td>Yes</td>
<td>Yes</td>
<td>10</td>
<td>2</td>
</tr>
</tbody>
</table>

TFC = Total Functional Capacity, NA = Not applicable.
end-of-life wishes (Table 3.2, Quotations 1 and 2). End-of-life wishes usually consist of thoughts concerning euthanasia or PAS in the course of the disease. Euthanasia, end-of-life wishes and the threshold of what an acceptable life or what quality of life entails are concepts that are intertwined when thinking and talking about this subject.

### Table 3.2: Background and presence of end-of-life wishes in HD patients and conversations with their physician about the wishes

<table>
<thead>
<tr>
<th>Quotation</th>
<th>Translation</th>
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</thead>
<tbody>
<tr>
<td>1 R11: “When I saw my mother in the end stage of HD, I thought, if I have to go through this one day, will I be able to cope.”</td>
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<tr>
<td>2 R9: “Not the way my mother suffered.”</td>
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<tr>
<td>3 I: “Do you ever think about the future?” R4: “Of course, every day.” I: “what do you think? Are there things you would want?” R4: “I am not going to leave home. I have seen that with my mother. As soon as I become dependent on others, then it is time to pull the plug. When I am not able to eat independently or be independent, that is unacceptable. If my quality of life diminishes to a point that I become dependent, than I quit.”</td>
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<tr>
<td>4 I: “Do you have any wishes or ideas for the future?” R9: “Well yes. That you tell others that you don’t want to go on. That you tell about what is acceptable and what is not, you know. Say, if you can’t go to the toilet anymore.” I: “Did you ever think about what would be acceptable for you and what not?” R9: “Well, if I have to move to a nursing home, that seems awful. Because I can’t take my cats with me.”</td>
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<td>5 I: “And if I ask about that, about the end of life. Is that something to discuss?” R1: “Yes, we can discuss that.” I: “What does it mean for you?” R1: “If I have to be fed, then, I think, I will have had it.”</td>
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<tr>
<td>6 R1: “We wondered sometimes if it is at all possible. We didn’t know if it might be possible to make arrangements now or in fact whether it is possible to make arrangements at all.”</td>
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<td>7 I: “You said to me that when things become worse, that would not be preferable. You talked about suicide. Did you ever think about euthanasia?” R8: “Yes, euthanasia, yes, but to come to that point, things have to be a lot worse, my situation would have to get worse. If I look at my dad, he seems content with the situation. But with me, things can be totally different.”</td>
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<td>8 R11: “Every day I keep hoping that the solution in Canada, with mice, where the disease has been halted. I have hopes that maybe a time will come for me, when I am not that ill, that I have to start thinking about euthanasia; that a solution will be found in some other area.”</td>
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<tr>
<td>9 R3: “I had a conversation once with the NHP. About the future...I am glad you came. Because now I have told you what my wishes are.”</td>
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<tr>
<td>10 R2: “I do not want to make a decision about the future now. There are so many options nowadays. We will see.”</td>
<td></td>
</tr>
<tr>
<td>11 R7: “I usually keep it simple. It is easy said, but I am not going to make a fuss. What will be, will be.”</td>
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<tr>
<td>12 R13: “I do not see a long-term future for myself. I do not think I’ll have much of a say.”</td>
<td></td>
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<tr>
<td>13 R5: “I started thinking about euthanasia about 10 years ago. I saw another patient with HD who didn’t know anything and I thought that is not what I want. And I filled in an advance euthanasia directive. And now I am further down than I, eh, I said I did not want to move to a nursing home, but now here I am, I moved here a month and a half ago. Things were not going well at home. Then we talked about it, saying if it is like this, end it. And then things change, my wishes changed. And we got a granddaughter. So now I am in a nursing home.”</td>
<td></td>
</tr>
<tr>
<td>14 R1: “Of course you can’t say now how you will think in 10-15-30 years’ time.”</td>
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Presence of wishes

From the interviews, three patterns emerged regarding the presence of wishes. One pattern is recognised in a group of patients that has an advance directive or was making one at the time of the interview (4 respondents). The advance directive was usually a euthanasia directive and respondents talked about euthanasia and what their threshold of acceptable life would be (Table 3.2, Quotation 3).

The second pattern revealed patients who have some general ideas about their future perspective and mostly circumstances or events they wanted to avoid, but had not yet specifically articulated their intentions (5 respondents). They found it difficult to put their thoughts or wishes into words (Table 3.2, Quotations 4 and 5). Some of them wondered if it was possible already to document wishes or was it better to postpone (Table 3.2, Quotations 6 and 7). And sometimes a patient wanted to keep up hope and wait for a solution (Table 3.2, Quotation 8). The respondents in this group indicated that they would like to have their intentions known and/or documented, for the future (Table 3.2, Quotation 9).

The third group (5 respondents) has no (distinct) end-of-life wishes or questions regarding this subject, although the reasons behind this differ. Three patients take life as it comes. One of them had never really thought about the possibility of having a wish about the end of life (Table 3.2, Quotations 10-12). One patient had thoughts about death and being a burden to others, but these thoughts did not result in an end-of-life wish.

Response shift (adapting to the disease or deviating from anticipatory beliefs) is an issue of concern for all patients, those with no wishes, those with general ideas about their wishes and for patients with more specific wishes and advance directives. Some patients decided not to draw up an advance directive because of potential response shift (Table 3.2, Quotation 13-14).

Knowledge about euthanasia, advance directives and other end-of-life wishes

The majority of respondents have thoughts about the end of life and are aware of the concept of euthanasia. Most of them, however, do not know how to have their wishes respected and/or carried out. They were not able to say exactly what was required nor did they know what the concept of euthanasia or PAS really means, in terms of the law or in terms of requirements (Table 3.3, Quotation 1). On the other hand some already have
a fair grasp of the relevant underlying values of the requirements, without being able to make these explicit (Table 3.3, Quotations 2 and 3). Several patients thought that it is necessary to be able to talk about your wishes and ask for them to be carried out (Table 3.3, Quotation 4).

Role of family and physician

Family, partners and friends proved to be important for many patients. Their thoughts and wishes were usually discussed with these persons first, or patients indicated that they would want to talk to family and partner first (Table 3.4, Quotation 1). These discussions can be brief and not always directed towards the specific goal or intention of clarifying and discussing their wishes. The role of family members was sometimes even more important than the cooperation and role of their physician, usually the general practitioner (GP) (Table 3.4, Quotation 2). In fact not all patients had discussed their wishes with their physician in some cases ascribing this to the nature of their relationship with their physician, usually their GP. One patient discussed his wishes when he drafted an advance directive but not afterwards (Table 3.4, Quotations 3, 4 and 5). The patients who did talk about end-of-life wishes talked to their GP or nursing home physician (NHP).

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**Table 3.3** Knowledge about euthanasia, advance directives and other end-of-life wishes

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<table>
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<tbody>
<tr>
<td>1</td>
<td>I: “Can you tell me something about euthanasia?” R9: “That you write down your limit, so to say, what is acceptable and what is not, for example when you’re not able to shower anymore or when you need help to eat. And that you can end your life, when you reach that point.”</td>
</tr>
<tr>
<td>2</td>
<td>I: “we talked about euthanasia earlier, can you tell me what you know about euthanasia?” R8: “Euthanasia, You have to put it in writing in a euthanasia directive, that’s what you want. I think you have to talk to a doctor. It is not something that just passes. And it has to be a serious situation. That you suffer unbearably. But, that is different for everyone. That’s it.” I: “you said suffer unbearably, what would that behold?” R8: “Yes, it has to be that you do not see a way out for yourself. That you are in a certain situation that you are totally grasped by the disease and that things are very rough, to stay alive. To put it like this, yes.”</td>
</tr>
<tr>
<td>3</td>
<td>I: “you mentioned an advance euthanasia directive. Can you tell me how you became known with it?” R11: “Yes. I am not sure if I will be strong enough at that moment. I have always been strong, but maybe in the future I think that my life is not worth living anymore. Sometimes I think it is important to draw it up (advance directive), and sometimes I think, because you have to be totally ‘with-it’ before you sign, right? Yes, you have to be able to say that you can no longer cope with the situation and that these are the things you can no longer take.”</td>
</tr>
<tr>
<td>4</td>
<td>R1: “Often you are no longer allowed to make your own decision. When someone is too ill, it is not allowed anymore, someone else has to decide then.”</td>
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</table>
DISCUSSION

In The Netherlands, end-of-life wishes, especially euthanasia and PAS, are frequently discussed subjects. In a non-selected group of 19 HD patients who visited our clinic, more than half were willing to talk or had already talked about their disease perspective, including end-of-life wishes. Most of the wishes concern euthanasia or PAS, because of wanting a situation arising, and wanting to stay in control. But knowledge about the requirements concerning advance directives, euthanasia or PAS is sometimes limited and wishes remain vague.

Wishes

Earlier research showed that many patients in a nursing home (mean age >80 years) have end-of-life wishes; these may not always involve euthanasia or PAS. For many people, thoughts or wishes for the end of life consist of a general idea of what they want or want to avoid; in retrospect it is sometimes uncertain whether a person foresaw this particular situation, when making the advance directive. HD patients, usually much younger, know that their future will be characterized by deterioration and loss of control. Wishes are present, sometimes distinct, sometimes a vague indication of what they would and would not want. Some patients see their future as being too insecure to have wishes. And by expressing wishes or talking to your physician, the subject and the consequences become real; it can imply giving up hope as a patient.
In this study most patients frequently spoke about the adverse experience with an affected parent, which determined their wishes about end of life, sometimes leading to a wish for euthanasia. Their answers reflected a wish to stay in control of their own life and death; this would seem to be in accordance with the wish of patients with other diseases, such as cancer or ALS, but the background to these thoughts differs.\(^{12}\)

**Knowledge about advance directives and the role of family and physician**

Although we discovered that many patients in this study have end-of-life wishes, not all of them discussed these with their physician. Earlier research studies have reported that patients usually fill in their advance directive papers and discuss them briefly with their physician at the time, but not later.\(^{6}\) Little is known about the reason for the lack of discussion; we can draw several conclusions from our study.

Discussing end-of-life wishes requires a good and trusting relationship between patient and physician. Most patients referred to their GP when talking about end-of-life wishes, some to their NHP. Some patients hardly ever see their GP and do not take the initiative to discuss their wishes and thoughts, sometimes because the patient assumes his/her GP knows little about the disease. Moreover, patients lack knowledge about the value of an advance directive, the physician’s role and how to get to the point where the wishes are respected. This was demonstrated by the patient’s perceived importance of family in the process of drawing up an advance directive. In law however, the family is not part of the process. Family does not have to give their consent or agree with the patient. The conversation between patient and physician is the only thing that counts. A recent study in The Netherlands amongst Dutch citizens demonstrated an increased awareness about the option of euthanasia. It also revealed that citizens do not always use the terms of the law the way they are meant. In a vignette study, most citizens knew that family does not have to give consent, but agreed that family has an important role in the process.\(^{7}\)

Another indication of lack of knowledge is the observation that some HD patients seemed to think that things have been taken care of once the advance directive has been composed, discussed and filed. A recent case of euthanasia in a patient suffering from HD was deemed incorrect because of the very limited conversation of the patient with her physician about her wishes and threshold.\(^{10}\) A Dutch study showed that patients have high expectations of their advance directive in case of dementia while these expectations are not met by their physicians, usually attributable to the lack of a proper conversation about the topic.\(^{6,13}\)
Some 7% of the Dutch population has an advance directive in case of dementia; the focus on and attention paid by society to this subject shows an increasing awareness of the options offered by an advance directive, especially with the increase in prevalence of neurodegenerative diseases.\textsuperscript{6,23} But patients in the present study were not always aware that they could draw up an advance directive and express their wishes at an early stage of the disease.

Interestingly, a few months after the interviews we learned that two patients (Respondents 10 and 12) had in the meantime created an advance directive, drawn up together with their nursing home physician. It should be noted that we posed open questions and did not provide any information concerning Dutch euthanasia law. This shows that end-of-life issues are important subjects for patients. Patients may want to talk about the subject, but do not always know how, when or with whom. Taking this step and making arrangements for their own future gave both patients peace of mind, indicating the (apparent) importance of a physician asking actively about end-of-life wishes in all patients.\textsuperscript{24,25}

**Response shift, competence and the role of advance directives**

An important issue in the applicability of advance directives is response shift, especially in the context of a disease where cognitive decline is a key symptom.\textsuperscript{26-28} Twelve cases of euthanasia in patients in early stages of dementia were reported in The Netherlands in 2009.\textsuperscript{10} From a judicial point of view, it can be argued that the subject of response shift does not apply to patients with dementia because when patients become incompetent there is no longer any will, only their previous wishes remain judicially valid. Thus, it only matters if the physician knew his patient prior to the dementia. But this would entail totally disregarding the current situation, and the fact that patients sometimes do have an opinion about their present life. Promoting patient autonomy by applying a previous wish into an actual wish means ignoring the time between the writing and current events.\textsuperscript{29,30} GPs report they regard consistency of the will and physician-patient relationship as most important factors when deciding if response shift is an issue. NHP regard advance directives as a useful tool, but not a document on the basis of which they would make decisions.\textsuperscript{31} This supports the views from literature that it can be difficult to decide if an advance directive applies,\textsuperscript{4,32,33} but ignores the finding that early stage dementia patients are able to describe their situation\textsuperscript{27,29,34} or the fact that most HD patients have had a glimpse of their future, through a family member.
We explored the existence of end-of-life wishes in HD patients. Our questions were broad and directed towards thoughts about and wishes for the end of life in general and not immediately directed towards euthanasia. Most of the respondents talked about their wishes in the context of their threshold of acceptable life and euthanasia. This shows how much euthanasia and end-of-life wishes are intertwined in The Netherlands and that they are at the forefront of many patients’ minds.

We reached saturation but to substantiate our findings a quantitative study must be undertaken. One limitation is that we interviewed only two patients in the premanifest phase, although these did not reveal any difference in themes. Furthermore our group of patients represented all stages of the disease and ranged from before onset until being almost totally dependent, thus showing that disease stage did not alter the relevant themes.

A second possible limitation is the potential existence of cognitive deterioration in our group. Patients were not explicitly tested beforehand. But especially in a disease where cognitive deterioration is a key symptom, we think it is important to ask these patients in particular for their opinion and ask about their wishes. The study showed that most patients are still able to speak about their lives and their wishes and that it was possible to extract these. Thirdly, the interviews were carried out by the first author of this article, who also is a physician. This fact was known to the patients. Bias is possible because of the role of the researcher, especially with a subject where the physician is the central figure in Dutch law. But the author was not the treating physician and we emphasized that the opinions would be dealt with confidentially and not provided to a GP or other treating physicians.

To the best of our knowledge, this is the first study in which patients were actively asked for their thoughts and attitudes towards the end of life and the content of wishes for the end of life, without prior selection. The study demonstrated that asking patients these questions is not problematic.

Patients with HD think about their future perspectives and how to stay in control of both their life and their death. The example of a parent is the strongest motive for having end-of-life wishes and determining a threshold of acceptable life. Probably more patients have wishes than those known to their physician, but have not communicated these with their physician for several reasons, including a lack of knowledge. Both the finding that patients were not always aware of the fact that it is possible to draw up an advance directive at
an early stage of the disease and the finding that only a minority of individuals with early dementia are able to complete an advance directive points out the important conclusions of an earlier study on advance directives in HD that it is important to talk about this subject early on in the disease.\textsuperscript{35,36}

If such a conversation is initiated by the physician, response shift can be detected at an early stage, possibly preventing anxiety because of future incompetence. For patients, more information about the (im)possibilities of an advance directive can help them put their wishes into words so that these are carried out in good time. Future research is needed to study if the presence of wishes and the content of these wishes is associated with certain patient characteristics, such as age, gender, religion or disease stage.

Not only for patients in the Netherlands, but also for patients in other countries where advance treatment directives are valid, possibly not for euthanasia, but for other medical decisions, these conclusions may help physicians to engage in conversation, provide information, increase knowledge and inform a patient in good time about their options and about how to ensure their wishes are respected. And to extend this suggestion even further, these conclusions could also help physicians in countries where advance directives do not have a status to realize the possibility that thoughts and wishes for the end of life might be present. Furthermore these conclusions can be applied not only to HD, but also more widely to other neurodegenerative diseases.

**Acknowledgements**

We thank all patients for their participation in this study. We thank Brenda Vollers for her help with the English.
REFERENCES


