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# Wishes for the end of life in Huntington's Disease

## Observations and reflections, initiated in The Netherlands

**Suzanne J. Booij**

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# Wishes for the end of life in Huntington's Disease

## Observations and reflections, initiated in The Netherlands

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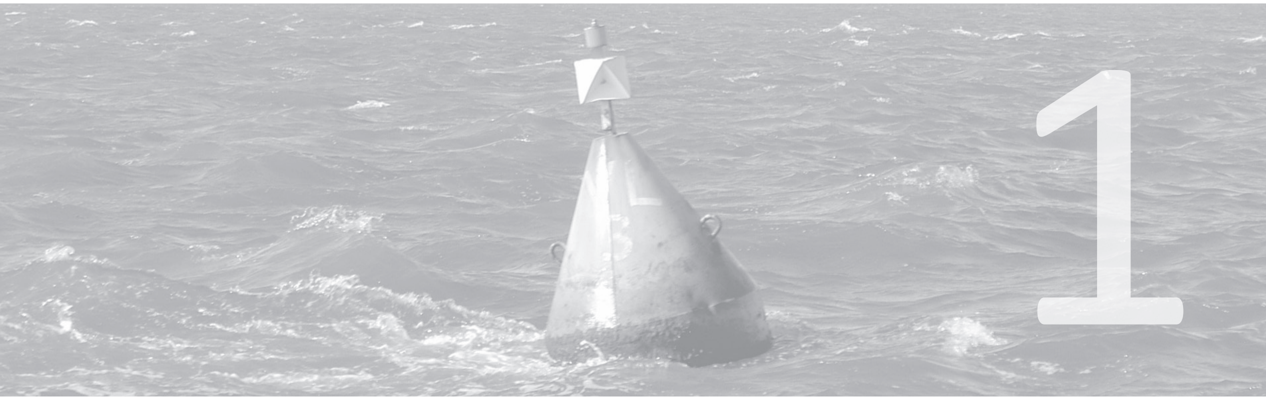


# 1

# Introduction and aims

In 1872 George Huntington described an illness characterized by chorea and psychiatric symptoms with a hereditary nature. This illness is since called Huntington's chorea and in the eighties of the last century Huntington's disease (HD).[1] Huntington's disease is characterized by a triad of psychiatric, motor and cognitive symptoms. In clinical practice most patients are diagnosed when motor symptoms appear, although the other symptoms may precede. The genetic mutation that causes HD was discovered in 1993 and since then predictive testing with a theoretical 100% accuracy is possible. The *HTT* gene defect, an elongated CAG repeat, is localized on chromosome 4.[2] Huntington's disease is a neurodegenerative disorder and is autosomal dominantly inherited. This means every child of an affected parent has a 50% chance of inheriting the gene and thus the disease. The option of predictive testing for HD by direct mutation analysis has been available for at-risk individuals since the CAG repeat expansion was identified. Main reasons for requesting the test are relief from uncertainty and planning for the future, with the inclusion of exerting control over the end stages of the disease. Uptake of predictive testing is consistently reported in between 5% and 25% of individuals at risk. Next to predictive testing, prenatal or preimplantation genetic diagnosis for HD is available in The Netherlands.[3]

Symptoms and signs of HD typically become manifest between the age of 30 and 50, affecting relatively young people, who usually still have a job and a young family. Disease duration is approximately 17-20 years. All patients will become totally dependent for all daily life activity and usually need nursing home care during the last couple of years.[4;5]

The existence of the disease within families, usually over many generations, has resulted in extensive knowledge of the course of the disease for family members at risk. After predictive testing or clinical diagnosis many patients know what to expect in the future. This knowledge and the possibilities to make choices and the need for control regarding the end of life, including the options of euthanasia and physician assisted suicide (PAS), resulted in an increased awareness amongst patients with HD, their family members and physicians that this is an increasingly important topic.

## Termination of Life on Request and Assisted Suicide Act in The Netherlands

The debate about euthanasia started in the 1970s when awareness in society grew that sometimes physicians support patients in the dying process.[6] During that same period important political developments had taken place. The Royal Dutch Medical Association

(KNMG) was asked about their opinion on euthanasia along with several other organizations. The KNMG did not take a position for or against euthanasia but propagated the view that the medical profession was prepared to take responsibility for euthanasia.[6]

In 2002 the process resulted in the “Termination of Life on Request and Assisted Suicide Act”, which codified the requirements of due care and made the Regional Review Committees (RRCs) principally responsible for reviewing reported cases.[6]

**Table 1.1** The requirements of due care

| | |
|---|---|
| 1 | The physician is convinced that the patient’s request was voluntary and well considered |
| 2 | The physicians in convinced that the patient’s suffering was unbearable and there was no prospect of improvement |
| 3 | The physician informed the patient about his situation and prospects |
| 4 | The physician and patient were convinced that there was no reasonable alternative in light of the patient’s situation |
| 4 | The physician consulted at least one other, independent physician, who must have seen the patient and given a written opinion on the requirements of due care |
| 5 | The physician terminated the patient’s life or provided assistance with suicide with due medical care and attention |

Since the codification, euthanasia and PAS have been subject of ongoing political and societal debate. Nevertheless in the course of these almost 30 years the public opinion also evolved to the point that a good 90% of the population is not against the legalization of euthanasia.[6]

Since the law of 2002 passed each year more reports on euthanasia and/or PAS are reported to the RRCs (Figure 1.1).[7] In 2012 4,188 persons died as a result f euthanasia or PAS, which represent approximately 3% of all deaths in The Netherlands.[7;8]

The possibility of euthanasia based on an advance directive is codified in article 2, paragraph 2 of the law. This paragraph of the law states that in case of an advance directive, the advance directive can replace the direct (oral) request for euthanasia. In this case the other requirements of due care are accordingly applicable and should be met.

From 2008 and onwards the RRCs received reports of euthanasia in case of dementia. Common policy of the RRCs is to handle these reported cases with care and caution, because of the nature of the disease, the nature of the request and the consequences of the request.[7]

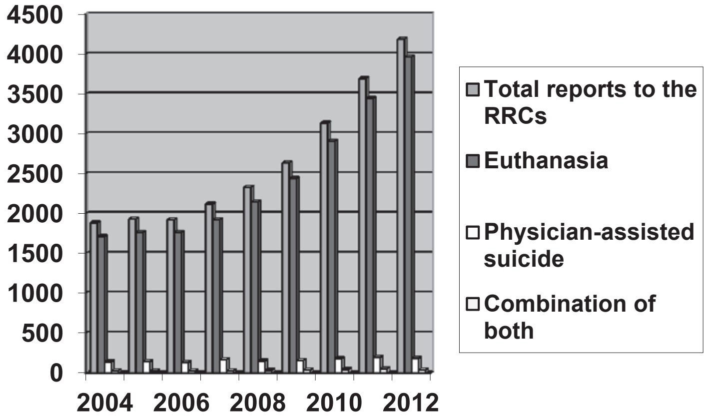


**Figure 1.1** Number of persons who died by euthanasia and PAS since 2004.

## End of life clinics

With an increased attention for the possibilities of euthanasia based on an advance directives and in case of dementia, the debate seems to shift towards having end-of-life wishes and the value of advance directives in neurodegenerative diseases, such as Alzheimer's disease or Huntington`s disease especially in advanced stages of these diseases.[9;10]

The increased attention has come to a new peak in The Netherlands with the establishment of end-of-life clinics in 2012. The aim of these clinics was to enable patients, who could not find their own physician willing to perform euthanasia or PAS, to die peacefully as a result of euthanasia or PAS, on the condition that the patient fulfills all the requirements of due care of the Euthanasia Act. These clinics consist of specialized teams of a physician and a nurse. First, members of the clinic perform an assessment on paper and decide if the request seems legitimate. If a request seems legitimate and feasible a team further investigates the request and contacts the patient. The team then decides if the request meets the requirements of due care according to the law. In the first 18 months since the advent of the clinics (in August 2013) 1,064 patients approached the end-of-life clinic in order to ask for euthanasia or PAS. About 10 percent of the patients died as a result of euthanasia, administered by a team from the clinic; another almost 10% found their general practitioner willing to perform the euthanasia in the end, usually after mediation of a team of the clinic. Furthermore, about 10% of the patients decided to withdraw their request. Fifteen percent of patients died before the request could be further investigated or fulfilled and about 23% of patients await the help of the teams of the end-of-life clinics. In 29 percent of cases the request was denied by the clinic, for two main reasons. One reason was administrative, for example the refusal of the patient to obtain his medical file with his/her treating physicians and the second reason was that the medical team of the clinic could not perform euthanasia or PAS because the patient did not meet all the requirements of due care.[11]

## Termination of Life on Request or Assisted Suicide Acts: world wide

An increased attention for the topic of euthanasia and assisted suicide emerged in other countries as well. End-of-life wishes became part of the debate in health care and part of the debate about determining and keeping quality of life. This attention resulted in legislation in some countries in respond to the increased questions from patients, families and health care workers.[6]

First attempts to come to codification of the subject of euthanasia and PAS were done in Oregon, USA in 1994. In 1994 a law passed making PAS legal, but euthanasia illegal. The law became effective in 1997 due to legal challenges. PAS is allowed in case of a terminal illness, when a patient is older than 18 and has less than 6 months to live. A written request must be present, signed by two witnesses. In 2008 and 2009 two other States, Washington and Montana, legalized physician assisted suicide. Euthanasia is still considered a crime.[12;13]

In Switzerland assisted suicide has been legal since 1918. The law does not require assistance from a physician and motives for the law were not primarily medical.[12;13]

In Belgium studies showed that euthanasia and PAS were executed without judicial confirmation. For this reason in 2002 the Belgian parliament enacted a law to legalize euthanasia and PAS, either after a direct request or based on an advance directive, after the requirements of due care have been met.[6]

The last country to legalize euthanasia in recent history was Luxemburg. Although the law had been in preparation for many years, it was not until 2009 that parliament passed the law. The delay was incurred by the refusal of the grand duke of Luxemburg, based on his religious beliefs, to sign the law. In response parliament took away some of the powers of the grand duke, making it not necessary anymore that the grand duke should sign all laws.

Nowadays terminally ill patients can request euthanasia after consent of two physicians and a panel of experts. Other requirements of due care resemble those required in The Netherlands.[14]

## Aims of this thesis and research questions

Most patients in The Netherlands will discuss their wishes for the end of life with their general practitioner, but for HD patients, both nursing home physicians and medical specialists, neurologists and psychiatrists, will be involved early during their disease course and thus in a conversation about the subject of wishes for the end of life. Clinical experience learns that patients with HD and identified mutation carriers show interest in the options of euthanasia and PAS. We observed in our out-patient clinic that an increasing number of patients brought the subject under the attention of the physician. Therefore, we wished to investigate what, how and when this was actually the case.

In the first chapter we investigated the theoretical option of euthanasia in HD, after a direct request or based on an advance directive (**chapter 2**).

To investigate whether HD patients or identified mutation carriers actually have end-of-life thoughts or wishes and whether they are willing to talk about these wishes we conducted two studies, one qualitative and one quantitative study. If end-of-life thoughts and wishes are present, we wanted to explore the content of these wishes and whether a patient seeks information or finds out how to have these wishes documented and respected. Secondly we want to study whether there actually is an increase in questions and deliberations concerning these issues (**chapter 3 and 4**).

We also wanted to study the perspective of physicians when talking and thinking about euthanasia and/or PAS in case of HD. Do physicians actually discuss this topic more often and do they receive questions concerning the topic of end-of-life wishes, both in The Netherlands and other European countries and how do physicians cope with and react to these kinds of questions from HD patients (**chapter 5, 6 and 7**).

In the final chapter (**chapter 8**) the principal results of this thesis are put together. They provide a starting point for both physicians treating patients with other neurodegenerative diseases and patients suffering from these diseases to enhance treatment and seek for the possibility to not only talk about quality of life, but when talking about quality of life in such a case, also about quality of dying.

# REFERENCES

(1) Huntington G. On Chorea. Med Surg Rep 1872;26:317-21.

(2) A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington's disease chromosomes. The Huntington's Disease Collaborative Research Group. Cell 1993 Mar 26;72(6):971-83.

(3) Tibben A. Predictive testing for Huntington's disease. Brain Res Bull 2007 Apr 30;72(2-3):165-71.

(4) Roos RA. Huntington's disease: a clinical review. Orphanet J Rare Dis 2010;5(1):40.

(5) Sturrock A, Leavitt BR. The clinical and genetic features of Huntington disease. J Geriatr Psychiatry Neurol 2010 Dec;23(4):243-59.

(6) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(7) http://www.euthanasiecommissie.nl, 2014.

(8) Onwuteaka-Philipsen BD, Brinkman-Stoppelenburg A, Penning C, de Jong-Krul GJ, van Delden JJ, van der Heide A. Trends in end-of-life practices before and after the enactment of the euthanasia law in the Netherlands from 1990 to 2010: a repeated cross-sectional survey. Lancet 2012 Sep 8;380(9845):908-15.

(9) Rurup ML, Onwuteaka-Philipsen BD, van der HA, van der WG, van der Maas PJ. Physicians' experiences with demented patients with advance euthanasia directives in the Netherlands. J Am Geriatr Soc 2005 Jul;53(7):1138-44.

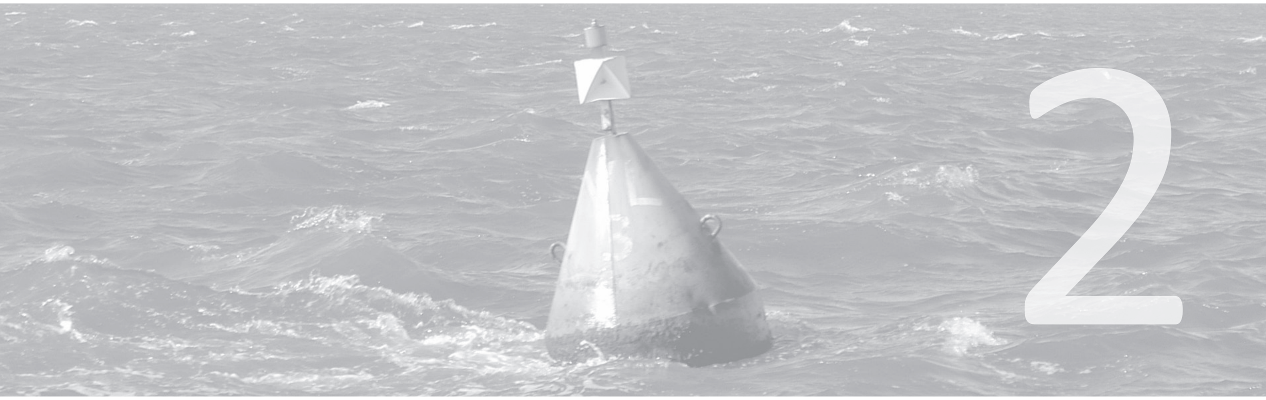
(10) van Delden JJ, van der Heide A, van de Vathorst S, Weyers H, van Tol DG. Kennis en opvattingen van publiek en professionals over medische besluitvorming en behandeling rond het einde van het leven. Het KOPPEL-onderzoek. ZonMw, Den Haag; 2011.

(11) www.levenseindekliniek.nl, 2014.

(12) Lewis P, Black I. Adherence to the Request Criterion in Jurisdictions Where Assisted Dying Is Lawful? A Review of the Criteria and Evidence in the Netherlands, Belgium, Oregon, and Switzerland. The Journal of Law, Medicine & Ethics 2013 Dec 1;41(4):885-98.

(13) Steck N, Egger M, Maessen M, Reisch T, Zwahlen M. Euthanasia and Assisted Suicide in Selected European Countries and US States: Systematic Literature Review. Med Care 2013;51(10).

(14) Watson R. Luxembourg is to allow euthanasia from 1 April. BMJ 2009;338:b1248.



# Euthanasia and physician-assisted suicide in Huntington's disease in the Netherlands

Suzanne J Booij
Dick P Engberts
Aad Tibben
Raymund AC Roos

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## ABSTRACT

Euthanasia or physician-assisted suicide is increasingly considered by patients with Huntington’s disease and identified gene carriers in The Netherlands. After codification of the Euthanasia Act in 2002 euthanasia performed by a physician is legal under strict conditions after a direct request from a competent patient and/or on the basis of an advance directive. In the absence of any effective treatment for Huntington’s disease euthanasia or physician-assisted suicide can be an option for patients. The clinical, ethical and legal dilemmas associated with euthanasia or physician-assisted suicide in Huntington’s disease are discussed.

# INTRODUCTION

Huntington's disease (HD) is a progressive neurodegenerative disease, caused by an autosomal dominant inherited expansion of a CAG-repeat on chromosome 4, known as the huntingtin gene. The disease is characterised by chorea and hypokinesia, psychiatric symptoms and progressive cognitive decline leading to dementia. The onset of disease is usually between 30 and 50 years of age.[1] The duration of illness is between 10-20 years. The rate of suicide is higher compared to the general population.[2] Currently symptomatic treatment to alleviate the symptoms and signs is available, but there is no cure for this devastating disease. Because the disease is progressive, all patients eventually will become totally dependent for all daily life functions.[3]

The localization of the genetic cause in 1983[4] and the identification of the HD-gene in 1993 made premanifest DNA testing possible in individuals at risk.[5] Testing is performed after extensive information about the procedure and possible consequences according to internationally agreed guidelines.[1]

In the Netherlands the codification of the Euthanasia Act in 2002 made euthanasia legal under strict conditions. The public debate on the subject of euthanasia and dementia started in the early nineties and has been going on since then.[6] The law of 2002 regulated part of the subject, but did not end the debate.[7] The number of cases reported to the committee increased from 1815 in 2003 to 2636 in 2009.[8] Most (85%) euthanasia requests concern patients suffering from cancer. In the group of diseases of the nervous system most patients requesting euthanasia suffer from Amyotrophic Lateral Sclerosis (ALS) about which extensive research has been done.[9]

There are substantial differences between ALS and HD when considering end-of-life issues, which differences also apply to other neurodegenerative diseases. The mean duration of illness in ALS is 3 years, whereas in HD and most other neurodegenerative diseases the duration is much longer, over 15 years. A second difference is that cognition remains rather intact in ALS in contrary to HD and other neurodegenerative diseases. And a third argument is the fact that many HD patients and mutation carriers are knowledgeable of the characteristics and the course of the disease in the affected parent or other family members.

Besides clinical experience and anecdotal data there are no studies on euthanasia or physician assisted suicide (PAS) or other end-of-life questions in patients with HD. Some research has been done on euthanasia and advance directives in dementia in recent years in the Netherlands.[10] Considering the arguments mentioned above this is a field where research is warranted. In this article we search for the possibility of euthanasia or PAS and the role of advance directives in HD. This paper may serve the debate on euthanasia and PAS in HD.

## SUICIDE

After pneumonia suicide is the second most common cause of death in HD.[1] Two periods have been identified in which the risk for suicide is increased in HD. The first period is immediately before receiving a formal diagnosis of HD and the second period is when independence diminishes and symptoms and signs of the disease increase.[11]

Subtle cognitive deficits are present in pre-manifest patients. Also a higher prevalence of psychiatric disorders (depression and obsessive-compulsive disorder) in pre-manifest mutation carriers has been described compared to the general population.[12] Both symptoms might result in impaired judgement. Therefore one can speculate that suicide ideation could be classified as a psychiatric disorder, which is part of the signs of HD. But on the other hand the Dutch Association of Psychiatry stated in their guideline “Dealing with the request for euthanasia and/or PAS” that suicide is not always a sign of psychopathology.[13] When evaluating these two considerations on suicidal ideation, suicide can also be a well considered option for a patient or pre-manifest gene carrier to prevent deterioration and future dependence.[13] Similar motives have been mentioned in earlier research on reasons for euthanasia in elderly.[14]

## SHORT RECALL ON THE DEVELOPMENT OF THE EUTHANASIA DEBATE IN THE NETHERLANDS

Until the 1970s euthanasia and PAS were not debated in public in the Netherlands. The Penal Code holds killing as an offence, including killing on request or assisting suicide (Article 293 Penal Code).

In the 1970s these opinions slowly start to change when in Dutch society the awareness arrived that sometimes doctors support patients in the dying process. The word euthanasia enters the discussion, but the scope of the concept had yet to be determined.[6]

In the late 1980s it was decided that only active termination of life at the patients request is regarded as euthanasia in the Netherlands. In that same period euthanasia becomes accepted if performed by a physician under strict circumstances defined by courts and the Royal Dutch Medical Association. An effort to codify this practice had failed until 2002. The first euthanasia case to reach the Supreme Court was the case of Dr. Schoonheim (Table 2.1). This was the first time a physician was not found guilty after performing euthanasia.[15] In the following decade more cases were brought into court and the first steps towards codification were set.

**Table 2.1** Schoonheim case

| |
|---|
| In this case the general practitioner (GP) Schoonheim had performed euthanasia on a 95-year-old female who on several occasions had asked him to end her life. The patient was bedridden after a hip fracture and had deteriorating eyesight and hearing. Mentally she was in full possession of her capacities. After deliberating with the patient and her family, the GP acceded to her request. The Supreme Court considered that this situation could be a situation of necessity. The Court referred to the patient's unbearable suffering, the prospect of loss of personal dignity and to die in a dignified manner. The Court of Appeals then accepted his defence of necessity and acquitted the GP. |

## EUTHANASIA ACT IN THE NETHERLANDS

In the Netherlands the Euthanasia Act was approved by parliament in 2002. The law holds euthanasia and PAS as a criminal offence unless they are performed by a physician who acts according to six criteria and reports the case to a review committee (Table 2.2).

Regarding HD, physicians can encounter several problems when fulfilling these criteria for euthanasia or PAS. The six criteria are discussed below separately to see how they can apply to HD.

**Table 2.2** Six criteria of care for euthanasia[6]

| | |
|---|---|
| 1 | Request was voluntary and well considered. |
| 2 | Suffering was unbearable and without prospect or relief. |
| 3 | Patient must be informed about the situation and prospects. |
| 4 | No alternative solution for the situation. |
| 5 | One other physician must have seen the patient and given a written evaluation. |
| 6 | Ending of life must be performed professional and careful. |

## Explore the request from the patient, for the future or present time

### *Advance directives*

The introduction of the Euthanasia Act in The Netherlands made it possible to replace a request for euthanasia or PAS, which is intended to be executed in the near future, by an advance directive, which is intended to be executed in the future, as long as the other requirements are met. Now a competent patient can make a request in advance, to be executed when the patient becomes incompetent and loses his decision making capacities. With an advance directive patient's autonomy is respected and extended into the future.[16]

But an advance directive, like an oral request for euthanasia is not a right, but a request, without binding legal consequences.[6]

Although advance directives for euthanasia were originally largely intended to apply to patients with dementia, the validity of these advance directives is now debated in case of patients with dementia.[6;17] In patients with dementia the gradual progression allows them to adapt to the disease. This can make patients deviate from their anticipatory beliefs, called response shift.[16]

Decline or complete absence of disease-insight can be the cause of a change of opinion and patients can contradict their own advance directive.[7] Also patients can find it difficult to decide and specifically articulate in advance what the wishes will be. And when documenting the wishes, patients may not want to opt out future developments, for example new treatments or family circumstances.[18]

Thirdly Dutch studies on dementia and euthanasia showed that physicians aren't very inclined to follow the advance directive, because they think it is invalid or even illegal, the advance directive was not explicit enough or think euthanasia for a patient with dementia is unacceptable.[10;19]

But there is a difference between HD and patients with dementia regarding advance directives. Unlike dementia patients with HD usually know of the existence of HD in the family. Because most patients have had experience with HD they also usually have had many years to think about end of life, which can include euthanasia or PAS.

The elements of response shift, the accuracy of the will, future development in treatment and increasing incompetence make the draft of an advance directive difficult. But because of the knowledge of symptoms and signs, advance directives can be a good way of documenting at least the present will in HD.[7]

### *The premanifest or early manifest phase of the disease*

Once identified as gene carrier, individuals may have profound thoughts and worries about their future quality of life. For gene carriers in pre-manifest phase in our clinic we have observed an increase in conversations about end-of-life wishes, as part of their wish to exert more control over their future life. It is a challenge for the physician to seriously and empathically respond to the thoughts and wishes regarding end-of-life decisions. In this stage decision making capacities and competence are not affected yet. The physician has a task exploring the request when the patient discusses the advance directive. He has to be sure it is voluntary and well considered (Table 2.3).[20]

**Table 2.3** Recommendations to assess decision making capacity

| | |
|---|---|
| 1 | Be able to choose and express the choice. |
| 2 | Be able to understand relevant information. |
| 3 | Be able to reason about the situation and its possible consequences and to appreciate these consequences. |
| 4 | Be able to handle the information rationally and value it according to own morals and values. |

### *The manifest phase*

If the patient is in the advanced stage of the disease competence and decision making capacity is often a problem. There will usually be a diminished understanding and reasoning and expression of the will can lack consistency. The cognitive deterioration may be differently valued if the will is expressed to the physician for a longer period of time or has just recently been expressed.[6]

While exploring the medical decision making capacity studies have shown that patients with mild cognitive impairment already have a diminished understanding (of the information), reasoning about the situation, the choice and possible consequences.[21]

Although a recent study emphasizes that communication with elderly with dementia is in the early stages quite possible and essential for advance care planning to be adjusted to their actual wishes and needs.[22]

The decision making capacity and competency is assessed for the decision at hand and not for capacity and competence in general and must be distinguished from having a will.[20] There is still debate about whether the requested level of decision making capacity and competence of a patient depends on the gravity of the consequences of the decision. Most doctors feel that the graver the consequence, the more competence needed. But this view must be balanced against respecting patient autonomy.[23] For this reason different authors stated that it is not the solution that matters, but the way the patient comes to that solution.[24] In the case of Schoonheim the judiciary stated that when considering a request for euthanasia or PAS the physician has to take personality, intelligence and history of the patient into account.[15]

## Suffering

The second criterion in the Dutch Euthanasia Act poses merely definition differences. Communication seems essential to the assessment of suffering. But suffering is mainly psychological and is a subjective experience.[6;25]

In the pre- and early manifest phases of HD the suffering is not physical, but is mostly psychological and concerns the fear of future suffering due to loss of independence and dignity and change of personality. This fear is realistic and can be seen as unbearable suffering without prospect. The legislator saw ‘without prospect’ as when there are no adequate alternative management options for the disease, i.e. there are no ways to relieve, ease or undo the suffering.[25] Also after the hearing of Schoonheim, the court ruled that “the prospect of unbearable suffering, progressive deterioration and when knowing that dying with dignity is possible now, but most likely not in the future can qualify as suffering, taken the history, intelligence and personality of the patient into account.”[15]

In early manifest patients, symptoms such as depression or anxiety can be alleviated with medication to some extent. In the advanced stages the fear may have become reality. HD patients suffer from the decline in all functions in life, socially, professionally, and loosing independence, and often with full awareness.[10] Studies with patients in the early stages of AD showed that patients are aware of their failing memory and are trying to find a way to cope with the problem. If these strategies work, chances of requests for euthanasia diminish and it is not sure if patients see their situation as suffering.[19;26] This is mainly part of the declining cognitive functioning and with that the decline of realization of cognitive deficits, the lack of awareness of being ill.[27]

But where in dementia the decline in cognitive functioning is the key aspect of the disease, in HD sometimes physical (marked hypokinesia and chorea) and psychiatric symptoms and signs precede the cognitive decline. In patients who encounter this course of the disease, the awareness of these signs and symptoms can be very present to them, and coping can be very difficult because there are little means to effectively treat these. For this reason HD patients can continue to see their situation as suffering and be aware of being ill. In this group chance of diminishing requests for euthanasia seem smaller.

## Information about situation and prospects

When considering euthanasia or PAS a physician is obligated to inform the patient about his situation, the possibilities of treatment and prognosis.[6] The information must be given at a time the patient is still competent and able to understand the information. Because HD is genetic and inherited, many have seen family members with HD and their signs and symptoms. Therefore patients may have had extensive information from other sources than their physician.[28]

## Alternative solutions

Pharmacological treatment allows alleviation of some of the symptoms and signs of HD, for example psychiatric signs and chorea. However the medication will not totally control the symptoms and signs which will become worse in due course.

In addition in the Netherlands chain care has been set up for HD patients. The chain care aims to take care of the patient from predictive testing until admittance to a nursing home. Even though admittance to a nursing home can give stability and symptoms can be relieved, it does not always provide a definite or desired solution for every patient. When offering chain care as an alternative solution it is important that the physician explores the frame of reference of his patient. Because relatives were usually taken care of in a different time with the absence of appropriate care it is possible that their reference is not up to date. The development of chain care shows that progress in HD care is made.

Statements preceding the euthanasia act stated that the alternative solution must be sought after extensive communication between physician and patient. There was no set time for this communication, so it can take place before the euthanasia request becomes pressing.[7] The decision to opt for euthanasia is a joint conclusion between physician and patient that for this problem, these symptoms there is no other solution left than euthanasia.[27] It is important as mentioned earlier to review the conclusions on a regular basis. A recent study emphasizes that communication with elderly with AD is in the early stages quite possible and essential for advance care planning to be adjusted to their actual wishes and needs.

### Consultation by an independent physician

In The Netherlands a system called Support and Consultation in Euthanasia in the Netherlands (SCEN) is available for consultation in euthanasia or PAS questions. These physicians are usually general practitioners (GP), are independent, specially trained, and available 7 days a week for consultation.[29] The consultation of an independent physician is obligatory and SCEN offers a network that meets the requirements.

### Ending of life is performed professionally and carefully

There are protocols in which way to perform euthanasia or PAS. Especially regarding the medication used and the obligatory presence of a physician in the case of PAS this sometimes leads to problems. This is mostly due to a lack of knowledge with the physician.[29]

## ANALYSIS AND DISCUSSION, PRACTICE OF EUTHANASIA

In the Netherlands between 2007 and January 2011 a total of 30 HD patients died by euthanasia or PAS.[30] With an overall prevalence of 1/10,000 in the Netherlands there will be approximately 1,700 patients, and with a duration of illness of approximately 15 years, 100 patients will die each year and a 100 new patients will get a diagnosis of HD.[1] This means that, when approximately 7 patients die of euthanasia each year, 7% of patients suffering from HD die of euthanasia or PAS. In our clinic the number of requests from HD patients for euthanasia or PAS and advance directives concerning this issue seems increasing. This observation can either be a real increase or an increased awareness and interest in these issues and this observation can also reflect that both physicians and their patients are more willing and open to discuss end-of-life issues.

To date no case of euthanasia in a person with dementia based on an advance directive has been reported. In 2006 54% of physicians in the Netherlands said they will not perform euthanasia based on an advance directive in a patient with AD.[10] Rurup et al. found also that a large percentage of physicians in the Netherlands aren't familiar with the rules regarding euthanasia, and if they are familiar there are difficulties with interpretation of these rules.[10]

In 2009 one case of euthanasia based on an advance directive was performed on a patient suffering from severe aphasia. In the years before the patient and physician spoke frequently and extensively about euthanasia and under which circumstances the patient would feel that he was suffering to such an extent that euthanasia was considered an option. The advance directive was clear and the frequent conversations with the physician made that the physician was very well informed about the wishes of the patient. Euthanasia was performed and declared accurate and careful by the regional review committee. The physician stated that not performing euthanasia would be abandoning the patient. It would be disrespecting patient autonomy to leave him in a situation he wished to avoid and which he considered as unbearable suffering.[8]

This line of reasoning of the physician and the acceptance by the regional review committee provides a clear example of the possibilities of euthanasia or PAS in HD. Especially the pre-manifest phase can be appropriate for physician and patient to explore and articulate the wishes and draw an advance directive. As we can deduct from the arguments mentioned before in the discussion of the criteria, the option for euthanasia or PAS in HD is within the scope of the law.

Further clinical knowledge is needed to find the reasons for asking for euthanasia by the patient and knowing the reasons for performing or not performing euthanasia as a physician. Also further clinical knowledge is needed to determine the role of advance directives.

When there is no cure and there are little means to treat and/or diminish suffering, discussing end-of-life wishes can also be a way of guidance and treatment of your patient.

## REFERENCES

(1) Roos RA. Huntington’s disease: a clinical review. Orphanet J Rare Dis 2010;5(1):40.

(2) Bindler L, Travers D, Millet B. [Suicide in Huntington’s disease: a review]. Rev Med Suisse 2009 Mar 18;5(195):646-8.

(3) Bates G, Harper PS, Jones L. Huntington’s Disease. 2011. New York, Oxford University Press.

(4) Gusella JF, Wexler NS, Conneally PM, Naylor SL, Anderson MA, Tanzi RE, et al. A polymorphic DNA marker genetically linked to Huntington’s disease. Nature 1983 Nov 17;306(5940):234-8.

(5) A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington’s disease chromosomes. The Huntington’s Disease Collaborative Research Group. Cell 1993 Mar 26;72(6):971-83.

(6) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(7) Hertogh CM, de Boer ME, Droes RM, Eefsting JA. Would we rather lose our life than lose our self? Lessons from the Dutch debate on euthanasia for patients with dementia. Am J Bioeth 2007 Apr;7(4):48-56.

(8) www.euthanasiecommissie.nl. Jaarverslag toetsingscommissie euthanasie 2009.

(9) Veldink JH, Wokke JH, van der WG, Vianney de Jong JM, van den Berg LH. Euthanasia and physician-assisted suicide among patients with amyotrophic lateral sclerosis in the Netherlands. N Engl J Med 2002 May 23;346(21):1638-44.

(10) Rurup ML, Onwuteaka-Philipsen BD, van der HA, van der WG, van der Maas PJ. Physicians’ experiences with demented patients with advance euthanasia directives in the Netherlands. J Am Geriatr Soc 2005 Jul;53(7):1138-44.

(11) Paulsen JS, Hoth KF, Nehl C, Stierman L. Critical periods of suicide risk in Huntington’s disease. Am J Psychiatry 2005 Apr;162(4):725-31.

(12) van Duijn E, Kingma EM, Timman R, Zitman FG, Tibben A, Roos RA, et al. Cross-sectional study on prevalences of psychiatric disorders in mutation carriers of Huntington’s disease compared with mutation-negative first-degree relatives. J Clin Psychiatry 2008 Nov;69(11):1804-10.

(13) www.nvvp.net; euthanasia and physician assisted suicide in psychiatric patients. 2014.

(14) Hardwig J. Is there a duty to die? Hastings Cent Rep 1997 Mar;27(2):34-42.

(15) Schoonheim case. Nederlandse Jurisprudentie 1984 Nov 27;(1985, 106).

(16) de Boer ME, Hertogh CM, Droes RM, Jonker C, Eefsting JA. Advance directives in dementia: issues of validity and effectiveness. Int Psychogeriatr 2010 Mar;22(2):201-8.

(17) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. Advance directives for euthanasia in dementia: do law-based opportunities lead to more euthanasia? Health Policy 2010 Dec;98(2-3):256-62.

(18) Lynn J, Teno J, Dresser R, Brock D, Nelson HL, Nelson JL, et al. Dementia and advance-care planning: perspectives from three countries on ethics and epidemiology. J Clin Ethics 1999;10(4):271-85.

(19) NVVE. Dementie en euthanasie. Er mag meer dan je denkt. [Dementia and euthanasia. More is allowed than you would think]. Amsterdam: NVVE, Nederlandse Vereniging voor een Vrijwillig Levenseinde; 2011.

(20) Moberg PJ, Rick JH. Decision-making capacity and competency in the elderly: a clinical and neuropsychological perspective. NeuroRehabilitation 2008;23(5):403-13.

(21) Okonkwo O, Griffith HR, Belue K, Lanza S, Zamrini EY, Harrell LE, et al. Medical decision-making capacity in patients with mild cognitive impairment. Neurology 2007 Oct 9;69(15):1528-35.

(22) van Delden JJ. The unfeasibility of requests for euthanasia in advance directives. J Med Ethics 2004 Oct;30(5):447-51.

(23) Buchanan A. Mental capacity, legal competence and consent to treatment. J R Soc Med 2004 Sep;97(9):415-20.

(24) Vellinga A. To know or not to be; Development of an instrument to assess decision-making capacity of cognitively impaired elderly patients. University of Amsterdam; 2008.

(25) Pans E. De normatieve grondslagen van het Nederlandse euthanasierecht. Nijmegen: Wolf Legal Publishers; 2006.

(26) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. [The lived-experiences of early-stage dementia and the feared suffering: an explorative survey]. Tijdschr Gerontol Geriatr 2010 Oct;41(5):194-203.

(27) Hertogh CM. The role of advance euthanasia directives as an aid to communication and shared decision-making in dementia. J Med Ethics 2009 Feb;35(2):100-3.

(28) Robins Wahlin TB. To know or not to know: a review of behaviour and suicidal ideation in preclinical Huntington’s disease. Patient Educ Couns 2007 Mar;65(3):279-87.

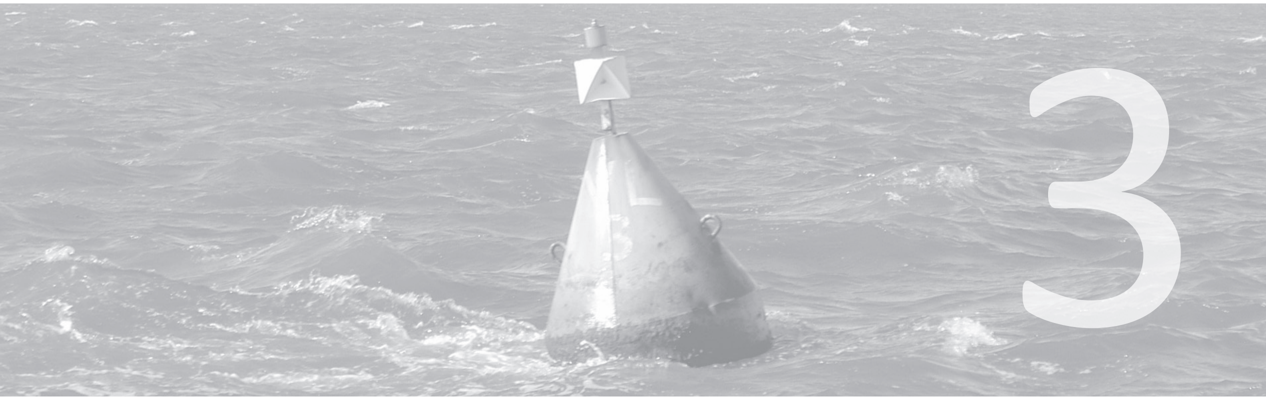
(29) www.knmg.artsennet.nl. SCEN. 2014.

(30) http://www.euthanasiecommissie.nl/contact/. 2014.

(34) de Boer ME, Hertogh CM, Droes RM, Riphagen II, Jonker C, Eefsting JA. Suffering from dementia - the patient's perspective: a review of the literature. Int Psychogeriatr 2007 Dec;19(6):1021-39.

(35) Bisson JI, Hampton V, Rosser A, Holm S. Developing a care pathway for advance decisions and powers of attorney: qualitative study. Br J Psychiatry 2009 Jan;194(1):55-61.

(36) Fazel S, Hope T, Jacoby R. Dementia, intelligence, and the competence to complete advance directives. Lancet 1999 Jul 3;354(9172):48.



# Euthanasia and advance directives in Huntington's disease: Qualitative analysis of interviews with patients

Suzanne J Booij
Verena Rödig
Dick P Engberts
Aad Tibben
Raymund AC Roos

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## 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.[1] 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,[1-3] the rate being 5-12 times higher in the HD population than in the general population.[3-5] 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.[6;7] 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.[8]

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.[9;10] The reason for most people in the Netherlands creating an advance euthanasia directive is to make arrangements in case dementia develops.[6] 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.[11;12] 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.[13;14] 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.[15]

In The Netherlands, between six and ten euthanasia requests from patients with HD were granted each year between 2007 and 2011.[10] 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.[16] 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.[17] 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[18] (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 3.1** Patient characteristics

| Patient/ respondent | Gender | Age | Marital status | Advance directive | Wishes | TFC | Disease stage |
|---|---|---|---|---|---|---|---|
| 1 | Female | 33 | Married | No | Yes | 13 | NA |
| 2 | Female | 37 | Married | No | No | 10 | 2 |
| 3 | Female | 54 | Relationship | No | Yes | 1 | 4 |
| 4 | Male | 43 | Married | Filling in papers | Yes | 11 | 1 |
| 5 | Male | 56 | Married | Yes | Yes | 1 | 4 |
| 6 | Male | 67 | Divorced, ex-wife care giver | Yes | Yes | 3 | 3 |
| 7 | Female | 68 | Married | No | No | 5 | 3 |
| 8 | Male | 44 | Married | No | Yes | 11 | 1 |
| 9 | Female | 28 | Single | No | Yes | 4 | 3 |
| 10 | Female | 45 | Relationship | No | No | 12 | 1 |
| 11 | Female | 35 | Relationship | No | Yes | 9 | 2 |
| 12 | Female | 70 | Single | No | No | 6 | 3 |
| 13 | Male | 44 | Married | No | No | 12 | NA |
| 14 | Male | 66 | Married | Yes | Yes | 10 | 2 |

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

| | |
|---|---|
| 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." |
| 2 | R9: "Not the way my mother suffered." |
| 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." |
| 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." |
| 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." |
| 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." |
| 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." |
| 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." |
| 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." |
| 10 | R2: "I do not want to make a decision about the future now. There are so many options nowadays. We will see." |
| 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." |
| 12 | R13: "I do not see a long-term future for myself. I do not think I'll have much of a say." |
| 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." |
| 14 | R1: "Of course you can't say now how you will think in 10-15-30 years' time." |

## 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

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

| | |
|---|---|
| 1 | 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.” |
| 2 | 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.” |
| 3 | 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.” |
| 4 | 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.” |

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).

**Table 3.4** Role of family and physician

| | |
|---|---|
| 1 | R6: "Yes, we are pro-euthanasia. I am preparing, together with my wife. The lawyer came and asked about an advance directive. And then we've gone back to our GP and we have the NVVE papers (Right to die-NL)." |
| 2 | R4: "My wife and friends have to give their consent. They have to make sure that my wishes are carried out if I can't see to it myself anymore." |
| 3 | I: "did you talk to your GP about your wishes?" R4: "No, not yet. Yes, he knows, I have the same GP as my mother had, so he knows about what should be done. But otherwise, I have no relationship with my GP. If I have any questions I visit the professor. And my GP, he doesn`t really know about the disease." |
| 4 | R7: "My GP, no I never visit him. No, he is nice, but he is of no help." |
| 5 | R5: "Yes, I talked about it with my GP and he gave me a leaflet about euthanasia. Then I handed in the advance directive. I do not remember what we talked about after that." |

## 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.[19] 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.[20] 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.[21;22]

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.[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.[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.[26-28] Twelve cases of euthanasia in patients in early stages of dementia were reported in The Netherlands in 2009.[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.[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.[31] This supports the views from literature that it can be difficult to decide if an advance directive applies,[4;32;33] but ignores the finding that early stage dementia patients are able to describe their situation[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.[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.

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## REFERENCES

(1) Bird TD. Outrageous fortune: the risk of suicide in genetic testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1289-92.

(2) Almqvist EW, Bloch M, Brinkman R, Craufurd D, Hayden MR. A worldwide assessment of the frequency of suicide, suicide attempts, or psychiatric hospitalization after predictive testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1293-304.

(3) Paulsen JS, Hoth KF, Nehl C, Stierman L. Critical periods of suicide risk in Huntington's disease. Am J Psychiatry 2005 Apr;162(4):725-31.

(4) Rurup ML, Onwuteaka-Philipsen BD, van der HA, van der WG, van der Maas PJ. Physicians' experiences with demented patients with advance euthanasia directives in the Netherlands. J Am Geriatr Soc 2005 Jul;53(7):1138-44.

(5) Farrer LA. Suicide and attempted suicide in Huntington disease: implications for preclinical testing of persons at risk. Am J Med Genet 1986 Jun;24(2):305-11.

(6) Rurup ML, Pasman HR, Onwuteaka-Philipsen BD. [Advance euthanasia directives in dementia rarely carried out. Qualitative study in physicians and patients]. Ned Tijdschr Geneeskd 2010;154:A1273.

(7) van Delden JJ, van der Heide A, van de Vathorst S, Weyers H, van Tol DG. Kennis en opvattingen van publiek en professionals over medische besluitvorming en behandeling rond het einde van het leven Het KOPPEL-onderzoek. ZonMW, Den Haag; 2011.

(8) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(9) van der Heide A, Onwuteaka-Philipsen BD, Rurup ML, Buiting HM, van Delden JJ, Hanssen-de Wolf JE, et al. End-of-life practices in the Netherlands under the Euthanasia Act. N Engl J Med 2007 May 10;356(19):1957-65.

(10) http://www.euthanasiecommissie.nl/contact/. 2011.

(11) Lindblad A, Juth N, Furst CJ, Lynoe N. Continuous deep sedation, physician-assisted suicide, and euthanasia in Huntington's disorder. Int J Palliat Nurs 2010 Nov;16(11):527-33.

(12) Maessen M, Veldink JH, van den Berg LH, Schouten HJ, van der WG, Onwuteaka-Philipsen BD. Requests for euthanasia: origin of suffering in ALS, heart failure, and cancer patients. J Neurol 2010 Jul;257(7):1192-8.

(13) Pasman HR, Rurup ML, Willems DL, Onwuteaka-Philipsen BD. Concept of unbearable suffering in context of ungranted requests for euthanasia: qualitative interviews with patients and physicians. BMJ 2009;339:b4362.

(14) Jansen-van der Weide MC, Onwuteaka-Philipsen BD, van der Wal G. Granted, undecided, withdrawn, and refused requests for euthanasia and physician-assisted suicide. Arch Intern Med 2005 Aug 8;165(15):1698-704.

(15) Onwuteaka-Philipsen BD, Gevers JK, van der Heide A, van Delden JJ, Pasman HR, Rietjens JA, et al. Evaluation of the law of review of ending of life on request and assisted suicide. ZonMW, Den Haag; 2007.

(16) Strauss AL, Corbin J. Basics of qualitative research: techniques and procedures for developing grounded theory. London: Sage Publications; 1998.

(17) Britten N. Qualitative interviews in medical research. BMJ 1995 Jul 22;311(6999):251-3.

(18) Shoulson I, Fahn S. Huntington disease: clinical care and evaluation. Neurology 1979 Jan;29(1):1-3.

(19) Kelner M. Activists and delegators: elderly patients’ preferences about control at the end of life. Soc Sci Med 1995 Aug;41(4):537-45.

(20) Lynn J, Teno J, Dresser R, Brock D, Nelson HL, Nelson JL, et al. Dementia and advance-care planning: perspectives from three countries on ethics and epidemiology. J Clin Ethics 1999;10(4):271-85.

(21) Ezzy D. Illness narratives: time, hope and HIV. Soc Sci Med 2000 Mar;50(5):605-17.

(22) Hines SC. Coping with uncertainties in advance care planning. J Commun 2001 Sep;51(3):498-513.

(23) Rurup ML, Onwuteaka-Philipsen BD, van der Heide A, van der Wal G, Deeg DJ. Frequency and determinants of advance directives concerning end-of-life care in The Netherlands. Soc Sci Med 2006 Mar;62(6):1552-63.

(24) Borgsteede SD, Deliens L, Graafland-Riedstra C, Francke AL, van der Wal G, Willems DL. Communication about euthanasia in general practice: opinions and experiences of patients and their general practitioners. Patient Educ Couns 2007 May;66(2):156-61.

(25) Dees MK, Vernooij-Dassen MJ, Dekkers WJ, Elwyn G, Vissers KC, van WC. Perspectives of decision-making in requests for euthanasia: A qualitative research among patients, relatives and treating physicians in the Netherlands. Palliat Med 2013 Jan;27(1):27-37.

(26) Hertogh CM, de Boer ME, Droes RM, Eefsting JA. Would we rather lose our life than lose our self? Lessons from the Dutch debate on euthanasia for patients with dementia. Am J Bioeth 2007 Apr;7(4):48-56.

(27) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. [The lived-experiences of early-stage dementia and the feared suffering: an explorative survey]. Tijdschr Gerontol Geriatr 2010 Oct;41(5):194-203.

(28) NVVE. [Dementia and euthanasia. More is allowed than you would think] NED: Dementie en euthanasie. Er mag meer dan je denkt. Amsterdam: NVVE, Nederlandse Vereniging voor een Vrijwillig Levenseinde; 2011.

(29) van Delden JJ. The unfeasibility of requests for euthanasia in advance directives. J Med Ethics 2004 Oct;30(5):447-51.

(30) Davis JK. The concept of precedent autonomy. Bioethics 2002 Apr;16(2):114-33.

(31) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. Advance directives for euthanasia in dementia: how do they affect resident care in Dutch nursing homes? Experiences of physicians and relatives. J Am Geriatr Soc 2011 Jun;59(6):989-96.

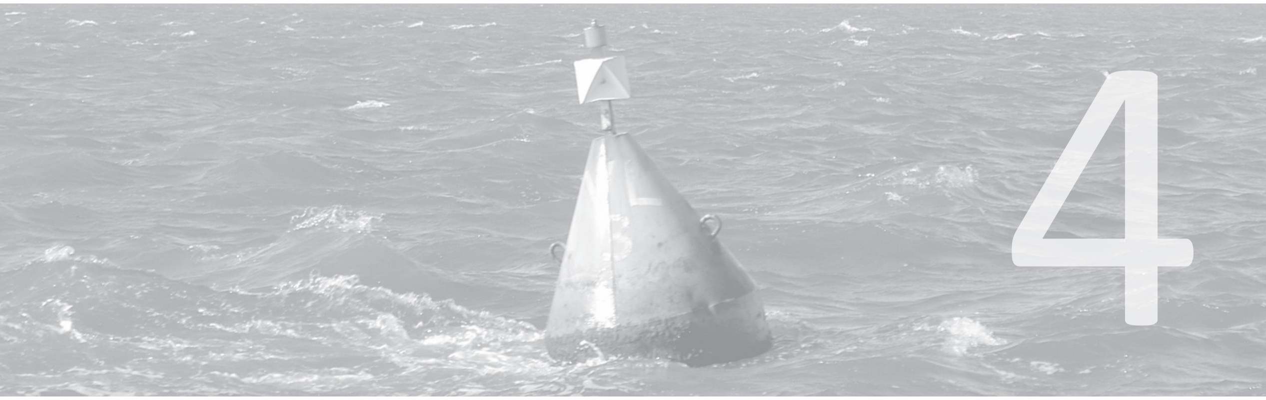
(32) Draper B, Peisah C, Snowdon J, Brodaty H. Early dementia diagnosis and the risk of suicide and euthanasia. Alzheimers Dement 2010 Jan;6(1):75-82.

(33) Gastmans C, De Lepeleire J. Living to the bitter end? A personalist approach to euthanasia in persons with severe dementia. Bioethics 2010 Feb;24(2):78-86.

(34) de Boer ME, Hertogh CM, Droes RM, Riphagen II, Jonker C, Eefsting JA. Suffering from dementia - the patient’s perspective: a review of the literature. Int Psychogeriatr 2007 Dec;19(6):1021-39.

(35) Bisson JI, Hampton V, Rosser A, Holm S. Developing a care pathway for advance decisions and powers of attorney: qualitative study. Br J Psychiatry 2009 Jan;194(1):55-61.

(36) Fazel S, Hope T, Jacoby R. Dementia, intelligence, and the competence to complete advance directives. Lancet 1999 Jul 3;354(9172):48.



# 4

# Thinking about the end of life: a common issue for patients with Huntington's Disease

Suzanne J Booij
Aad Tibben
Dick P Engberts
Johan Marinus
Raymund AC Roos

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## ABSTRACT

**Objective:** To investigate the presence of thoughts or wishes for the end of life in patients with Huntington's Disease (HD) or identified gene carriers (further mentioned together as patients).

**Methods:** A custom-made questionnaire, based on previous qualitative research, was sent out to 242 patients with HD and identified gene carriers. Presence of wishes was investigated and correlated to demographic and clinical characteristics.

**Results:** A total of 134 patients (55%) returned the questionnaire. One-hundred-and-one respondents (75%) reported to have some kind of thoughts or wishes for the end of life. For fifteen respondents (11%) these thoughts concerned care; eighty-six respondents (64%) reported to have also thoughts about euthanasia or physician assisted suicide (PAS). The presence of any thoughts about the end of life was significantly related to being familiar with HD in the family, but not related to any other demographic or clinical variable. Participants with thoughts specifically about euthanasia or PAS were of higher education and in earlier stages of the disease than participants without such thoughts.

**Conclusions:** Thoughts or wishes for the end of life are present amongst patients with HD. These thoughts include euthanasia or PAS in a majority of the respondents. It is suggested that prudential addressing these issues may enhance the doctor-patient relationship.

# INTRODUCTION

Euthanasia and physician assisted suicide (PAS) are legal in The Netherlands since the law of 2002 passed, but under strict conditions.[1] Euthanasia is defined as death brought upon by a physician at the patient's explicit request, either voiced orally or based upon a written advance directive.[2] Approximately 2% of all annual deaths in The Netherlands result from euthanasia or PAS.[3] In The Netherlands we recognize several kinds of advance directives. We recognize advance treatment directives, for example "Do not resuscitate" which a physician must abide to. A request for euthanasia or PAS, documented in a written advance directive is not a right of a patient but a request, without binding legal consequences.[4] Physicians generally do not initiate a conversation about end-of-life wishes and have difficulty handling an advance directive even though situations for which the advance directive was intended were recognized.[5-8]

Huntington's Disease (HD) is an inherited neurodegenerative disease, characterized by movement disorders such as chorea and hypokinesia, by cognitive decline leading to dementia, and by psychiatric symptoms. The localizing gene was identified in 1993 and this discovery made genetic testing possible.[9] In the Netherlands the prevalence is approximately 1,700 patients and another 6,000-8,000 people are at risk.[10] Between 50 and 60 patients suffering from HD die each year.[11] Since 2005 6-10 HD patients died every year after euthanasia or PAS.[12]

Qualitative research, by means of in-depth semi-structured interviews with physicians familiar with HD and HD patients preceded this study.[13;14] From these studies we concluded that most physicians leave it up to the patient to initiate a conversation about their wishes for the end of life. Furthermore we learned that some patients have specifically articulated their intentions, usually to family members. Other patients already had some ideas or wishes for the future, but did not know if it was possible to document these already or hesitated to discuss these, because their thoughts and wishes were not clear to them yet. We also found that patients underestimated the important role of their physician in the process of documenting wishes for the end of life and in the process of having these wishes fulfilled.

For the purpose of the present study we developed a questionnaire to explore the presence of end-of-life wishes, the reasons for having these wishes, the presence of advance directives and the conversations about end-of-life wishes in a larger group of HD patients or identified gene carriers (further mentioned together as patients) in the Netherlands. The first aim of this study was to explore whether there is any way to predict which patients are most likely to benefit from discussion of end of life issues. We wanted to explore if end-of-life wishes are present in the Dutch HD population, what the wishes include and if the presence of wishes is related to demographic or disease specific characteristics, such as cognition, quality of life and severity of motor symptoms. To investigate if wishes are related to any characteristics, we distinguish two groups, a group of respondents with wishes and a group without any wishes. Furthermore, we make a (second) distinction between respondents considering euthanasia or PAS and respondents with other types of wishes. The second aim was to investigate if HD patients or identified gene carriers use advance directives and if they talk to family or a physician about their wishes.

## METHODS

### Study population

The study was a single center study in the Leiden University Medical Center, which is a national referral center for Huntington's Disease. Patients visiting our out-patient clinics are requested to register in the International HD Registry. Those who consented to be included in the registry could also consent to being approached for participation in scientific studies. For this study we screened 296 patients that were included in the registry database since the database was set up. We screened between September and December 2011. Inclusion criteria were: a genetically proven status of HD gene carrier, age above 18, able to communicate either oral or in writing. Exclusion criteria were: no informed consent, suffering from severe depression according to the medical record, absence of a medical record, addiction to alcohol or drugs or having suicidal ideations at present. Fifty-five patients were excluded because they declined informed consent to be approached for scientific research, had severe depression or suicidal ideations or because their last visit was too long ago (>3 years) and we missed accurate information about their present condition.

In addition 5 patients were included via the Dutch Huntington's Disease patients' association after posting an announcement in their quarterly. Two hundred and forty two questionnaires were sent out in January 2012. A reminder was sent to non-responders in March 2012.

## Instruments and data collection procedure

A custom made questionnaire was developed for this study. The items were based on clinical experience, literature, and qualitative research that preceded this study.[15;16]

First, information on demographic characteristics was collected. Age was calculated on January 2012, when the questionnaire was sent out. Educational level was evaluated using a nine-point scale that was subsequently categorized into three categories, lower, middle and higher education (according to the categories used in the Registry European HD Network questionnaire). Data on the subject's religion was collected. The importance of religion for the patient was dichotomized into not important and important. Familiarity with HD was assessed by asking if the respondent was familiar with HD in the family and which parent is or was ill.

Second, to explore any wishes about the end of life, the first question concerned if respondents ever thought about the end of life and what the contents of these thoughts were. This question provided several options and the respondent could give multiple answers. In addition, reasons for having these wishes and conversations about the wishes, either with family members or with the general practitioners were assessed. Third, several questions assessed the presence of advance directives for end-of-life wishes. Items regarding the end of life in HD patients addressed euthanasia or PAS, tube feeding, admittance to a nursing home, care at home or treatment for other diseases. We categorized the responses to these items into no wishes, wishes except euthanasia or PAS and wishes including euthanasia or PAS.

Quality of life was evaluated using a nine-point numeric scale ranging from 1 (really bad) to 9 (excellent) (modified from SF-36[17]).

In addition to the custom-made questionnaire, global functioning was assessed using the Total Functioning Capacity (TFC) subscale of the Unified Huntington's Diseases Rating Scale (UHDRS). The TFC consists of 5 questions assessing employment, the capacity to handle financial affairs, manage domestic chores and perform activities of daily living; and the care level provided.[18] Motor function was assessed using the UHDRS motor score (UHDRS-M), ranging from 0-124 points.[19] A score below 5 denotes no motor abnormalities. Mini-Mental State Examination (MMSE) data were collected to assess global cognitive functioning.[20] The TFC and UHDRS-M are performed by a neurologist and the MMSE is performed by a neuropsychologist, both experienced with HD, at every visit to our out-patient clinic as part of the Registry data collection. The TFC, UHDRS and

MMSE scores used in the present study were those that were obtained at the time point closest to the date the questionnaire was completed (or sent), and had to have been collected within the previous 12 months. If the last available test score was more than one year earlier, the result was recorded as missing.

### Statistical analysis

Data were analyzed with SPSS 20.0. Data are presented as n (%) or mean (SD) as appropriate. The Chi-square test (or Fisher's Exact Test, when appropriate) was used to test for group differences with respect to categorical data, whereas t-tests for independent samples were used for normally distributed continuous data and Mann-Whitney U tests for samples with non-normally distributed continuous data. The significance level was set at $p < 0.05$.

## RESULTS

### General respondent information

One-hundred-and-thirty-four out of 242 questionnaires were returned, a response rate of 55.4%. One respondent returned the questionnaire with negative remarks, indicating that this was a subject the respondent did not want to discuss (Figure 4.1). If a respondent decided not to answer to a specific question, the result was recorded as missing. Respondent and non-responder characteristics are summarized in Table 4.1. The non-responders that were deceased or moved were not included in the analysis.

Thirty-four respondents have the maximum TFC score of 13. Thirty-one respondents have the maximum MMSE score of 30 and 36 respondents have a motor score of 5 or lower.

Non responders were younger and of lower education than responders, but were not different considering gender, having a partner, or disease specific characteristics such as TFC and UHDRS-M.

### Thoughts about the end of life

Thirty respondents indicated to have no thoughts about or wishes concerning the end of life. Three respondents did not answer this question. The remaining 101 respondents

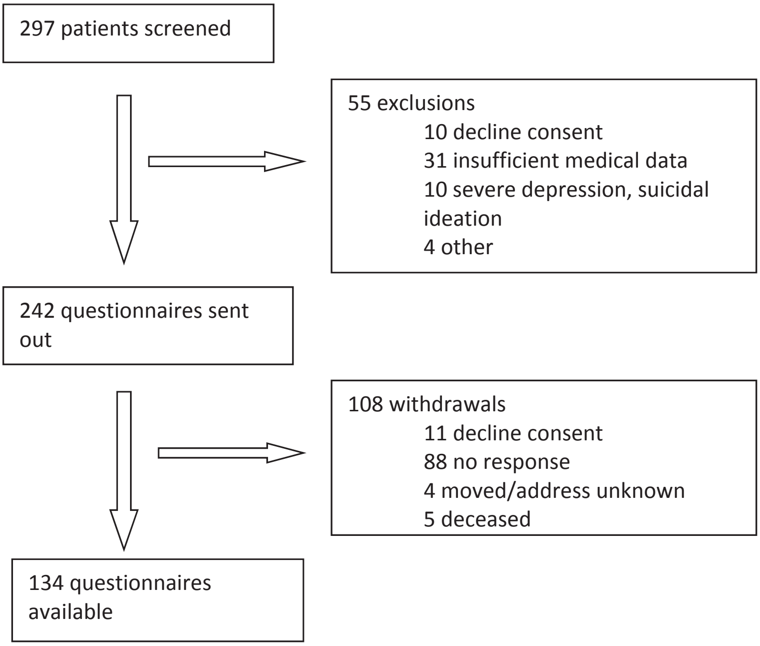

**Figure 4.1** Study profile.

**Table 4.1** Characteristics of 134 respondents and 99 non-responders

| | Respondents | Non-responders | p-value |
|---|---|---|---|
| **Demographics** | | | |
| Female | 71 (53%) | 63 (64%) | 0.06 |
| Age, years (mean, range) | 51 (21-85) | 47 (20-78) | 0.02 |
| Married or partner | 100 (74.6%) | 63 (61.8%) | 0.06 |
| Education | | | 0.001 |
| Lower level | 35 (26.1%) | 35 (34.0%) | |
| Middle level | 52 (38.8%) | 51 (49.5%) | |
| Higher level | 46 (34.3%) | 14 (13.6%) | |
| Religious | 43 (32.1%) | - | |
| Acquainted with HD in family | 101 (75.4%) | - | |
| Conversation with family about HD | 125 (93.3%) | - | |
| **Clinical characteristics** | | | |
| TFC (mean, SD) | 8.5 (3.9) | 8.3 (4.3) | 0.72 |
| UHDRS-M (mean, SD) | 30.2 (26.1) | 28.0 (27.4) | 0.57 |
| MMSE (mean) | 26.5 | | |
| Quality of life (mean, SD) | 6.02 (1.8) | - | |

HD = Huntington's Disease, TFC = Total Functional Capacity, UHDRS = Unified Huntington Disease Rating Scale-Motor, MMSE = Mini Mental State Examination.

indicated to have some thoughts about the end of life. Thoughts are for example: “Not the way my mother suffered” or “as soon as I become dependent on others, than it is time to pull the plug” or “euthanasia, yes, but to come to that point, things have to be a lot worse” (for further qualitative statements: see reference 14).

Having witnessed HD in close relatives was significantly associated with the presence of wishes for the end of life. The presence of wishes for the end of life was not associated with gender, age, marital status, education, considering oneself as a religious person, TFC, UHDRS-M, MMSE or quality of life (Table 4.2).

### Respondents with thoughts about or wishes for the end of life

Of the respondents who reported to have thought about the end of life, 86 indicated that wishes concerned thoughts about euthanasia or PAS together with other wishes for example about the possibilities of care. Fifteen respondents indicated that wishes concerned only the possibility of care, either at home or in a nursing home and possibilities of tube feeding and other medical treatments. The reason most often mentioned why thoughts about the end of life emerged was the loss of personal dignity.

**Table 4.2** Characteristics of patients with and without thoughts about the end of life

| | No thoughts (n=30) | Any thoughts (n=101) | Missing value | p-value |
|---|---|---|---|---|
| **Demographics** | | | | |
| Male | 12 (40%) | 49 (48.5%) | 0 | 0.41 |
| Age in years (mean, SD) | 47.1 (14.5) | 52.3 (13.7) | 0 | 0.08 |
| Married or partner | 22 (73.3%) | 77 (77.8%) | 1 | 0.68 |
| Education | | | 1 | 0.08 |
| Lower | 9 (30%) | 26 (26.0%) | | |
| Middle | 16 (53.3%) | 36 (35.0%) | | |
| Higher | 5 (16.7%) | 38 (38.0%) | | |
| Religious | 10 (33.3%) | 31 (31.0%) | 1 | 0.81 |
| Familiar with HD in family | 19 (63.3%) | 79 (81.4%) | 4 | 0.04 |
| **Clinical characteristics** | | | | |
| TFC (mean, SD) | 7.69 (4.5) | 8.69 (3.8) | 2 | 0.37 |
| UHDRS-M (mean, SD) | 31.07 (29.0) | 29.53 (25.4) | 2 | 0.94 |
| MMSE (mean, SD) | 25.58 (4.4) | 26.8 (4.2) | 12 | 0.14 |
| QOL (mean, SD) | 6.37 (1.7) | 5.97 (1.8) | 1 | 0.40 |

HD = Huntington’s Disease, TFC= Total Functional Capacity, UHDRS-M = Unified Huntington Disease Rating Scale-Motor, MMSE = Mini Mental State Examination, QOL = Quality of Life, SD = Standard Deviation.

Of the respondents who reported to have thought about the end of life 78 (77.2%) discussed these wishes with partner or family members. Forty-three (42.6%) respondents reported to have discussed their wishes with their general practitioner and 58 (57.4%) did not. Reasons for not talking about end of life wishes were not being ready for it in 26 (45%) of these respondents and the fact that they had little or no contact with their general practitioner in 16 (28%) respondents.

## Respondents considering euthanasia or physician assisted suicide

Eighty-six out of 134 respondents had wishes concerning euthanasia or PAS at some point in the disease process (Table 4.3). The respondents with wishes concerning euthanasia or PAS were of higher education and had lower motor scores, compared to the patients with wishes concerning only care. There was a trend towards higher overall functioning on the TFC for respondents who thought about euthanasia or PAS. Furthermore, respondents with wishes concerning euthanasia or PAS discussed their wishes more often with both

**Table 4.3** Characteristics of patients with end-of-life wishes

| | Wishes considering care (n=15) | Wishes considering euthanasia/PAS with or without care (n=86) | Missing value | p-value |
|---|---|---|---|---|
| **Demographics** | | | | |
| Male (n, %) | 6 (40%) | 43 (50%) | 0 | 0.48 |
| Age in years (mean, SD) | 56.1 (11.9) | 51.6 (14.0) | 0 | 0.25 |
| Active Religion (n, %) | 8 (53.3%) | 23 (27.1%) | 1 | 0.07 |
| Married or partner (n, %) | 13 (86.7%) | 64 (75.3%) | 1 | 0.51 |
| Education | | | 1 | 0.03 |
| Lower | 6 (40%) | 20 (23.5%) | | |
| Middle | 8 (53.3%) | 28 (32.9%) | | |
| Higher | 1 (6.7%) | 37 (43.5%) | | |
| Conversation with GP (n, %) | 1 (6.7%) | 42 (48.8%) | 0 | 0.002 |
| Good relationship with GP (n, %) | 11 (73.3%) | 72 (84.7%) | 1 | 0.28 |
| Familiar with HD in family (n, %) | 10 (66.7%) | 69 (84.1%) | 4 | 0.15 |
| Conversation with family | 7 (46.7%) | 71 (82.6%) | 0 | 0.005 |
| **Clinical characteristics** | | | | |
| TFC (mean, SD) | 7.1 (3.3) | 8.9 (3.8) | 1 | 0.06 |
| UHDRS-M (mean, SD) | 41.8 (24.9) | 27.4 (25.0) | 1 | 0.03 |
| MMSE (mean, SD) | 24.7 (5.3) | 27.1 (3.9) | 8 | 0.14 |
| QOL (mean, SD) | 5.3 (2.1) | 6.1 (1.8) | 1 | 0.19 |

GP = General Practitioner, TFC= Total Functional Capacity, MMSE = Mini Mental State Examination, UHDRS-M = Unified Huntington Disease Rating Scale-Motor, QOL = Quality of Life, SD = Standard Deviation.

family and their general practitioner than respondents with wishes concerning only care. There was a trend towards considering oneself more often as a religious person for respondents with wishes concerning only care at the end of life.

### Advance directives

Forty-two respondents (31.3%) had an advance directive; in 33 cases this is a euthanasia request, together with other kinds of advance directives, for example not to resuscitate (DNR) or a treatment prohibition, and in 8 cases this is a euthanasia request only. One respondent did not specify what kind of advance directive he/she had. Respondents with an advance directive are slightly older than respondents without, 54.6 years versus 49.2 years of age (p=0.038) and consider themselves more often as non-religious (p=0.029).

## DISCUSSION

This study shows that, when asked, the majority of HD patients visiting our out-patient clinic has thoughts about their end-of-life. The presence of wishes was not correlated with any demographic variable with the exception of familiarity with the disease in close relatives. The majority of HD patients did discuss their thoughts or wishes for the end of life with their family members, but less than half discussed these with a physician. The majority of the wishes consists of wishes concerning euthanasia or PAS. The presence of advance directives is limited.

This study shows that, at least on the basis of the variables included here, it is not possible for a physician to distinguish, in advance, between patients who have wishes and patients who do not. For this reason physicians should prudentially discuss wishes for the end of life. It is our experience that such attention for intimate issues may enhance and deepen the doctor-patient relationship.

### Presence of wishes

Seventy-one percent of respondents in a large Dutch study in the general population (n=1,980)  about end-of-life wishes indicated that they have thought about medical treatment and decision making for the end of life.[21] Mean age of the respondents was 53.7 years. Forty-one percent of respondents indicated that they had discussed this subject with family members, and only 4% with their physician. Respondents in that Dutch study were of higher education than in the general population.[21] Respondents in our study were about the same age. The percentage of respondents that have some kind of end of life wishes in our study is consistent with the national survey, indicating that in the Netherlands wishes for the end of life are present both in healthy citizens and in patients suffering from a neurodegenerative disease. We can conclude that patients with HD do not think about the end of life more or less frequent than persons from the general population, but that there are specific characteristics in HD patients that lead to these thoughts such as being familiar with the disease and motor signs and symptoms.

A larger percentage of respondents in our study discussed their wishes with either family members and/or their physician than in the general population. Most HD-families have witnessed the disease course in more than one generation and many of the family members face the disease themselves which may have raised ongoing discussions about what to do in the last stages of the disease.

Both respondents in the national survey and in our study were of higher education than non-responders. On the other hand, level of education was not correlated with the presence of wishes in our study and therefore it is unlikely that this potential source of bias influenced our results.

## Advance directives

In the Netherlands 95% of people know about the existence of a law concerning the end of life and 75% know what the term euthanasia means and includes.[21] Seven percent of Dutch inhabitants have an advance directive, usually a euthanasia request in case of dementia.[21;22] The percentage of patients with an advance directive is higher in our study sample than in the general population. This can be explained by the fact that we investigated a selected sample of known mutation carriers and HD patients. These patients with HD usually witnessed the appalling disease course in one or more close relatives. The need for control and have a say in their own future was reported as a reason for making an advance directive and considering specific arrangements for the end of life.[14] Staying in control of one's life and death and making one's own medical decisions is part of autonomy and is part of a patient's identity.[23;24] The fear of losing autonomy and dignity are described as reasons for asking for euthanasia or making other decisions for the end of life.[25;26] Research in the USA showed that the prevalence of advance directives increases with age or when people become ill. People seem to formulate an advance directive when it is likely they will need one.[27] Other research showed that discussing end-of-life choices by health care workers resulted in a selection of those choices by HD patients, relatives or legal representatives in 75% of individuals.[28]

## Religion

In our study considering oneself a religious person was not correlated with the absence or presence of wishes for the end of life, in contrast to other research where religious persons and patients have objections towards euthanasia.[29-31]

The Netherlands is a highly secularized country and the debate about euthanasia started in the 1960s.[4] The number of inhabitants visiting a church has been decreasing since 1960. Estimates are that around 2020 more than 70% of Dutch inhabitants will not be affiliated with any religion.[32] However in respondents who have wishes, there was a trend among religious patients to think more often about care than about euthanasia or PAS.

The strengths of this study are the relatively large study population of HD patients and identified gene carriers and the fact that we applied few exclusion criteria. There are some limitations that warrant discussion. First, the response rate was 55%. Analysis of non-responder characteristics showed that non responders are younger and of lower education, but their disease characteristics do not differ, thus indicating that age could be relevant but disease stage, gender and marital status are not; however, given that age was only marginally related to the presence of wishes, this seems unlikely. We can only speculate about the reasons for not filling in the questionnaire. It cannot be ruled out that particularly those patients who do not want to discuss end of life circumstances decided not to return their questionnaire, which may then have led to some overestimation. Additionally respondents of lower education may be less aware that they can already express their wishes without explanation from their physician first. Together this may have resulted in some selection bias and the study may thus not be regarded as representative for the Dutch HD patients. However, it may also be that non-response is not selective, given that the characteristics of non-responders in our population are similar to those in the general population, i.e. that they are also younger and less well educated. Unfortunately we have no information on the reasons for non-participation.

Second, the majority of respondents still live at home. The group of respondents that live in a nursing home and are in the latter stages of the disease is relatively small. Most patients who have been diagnosed with HD are still able to visit our out-patient clinic. Bias can occur because the wishes of relatively healthy respondents are included in this study and we have limited information of the wishes of more severely affected HD patients. On the other hand, consistent with studies in the USA, it is plausible that when patients become more severely affected and in possible need of an advance directive, and when patients and relatives are educated about the choices, the number of patients who think about the end of life and make choices, increases.

To our knowledge this is the first large study among HD patients to investigate end-of-life wishes and the first study where a relatively unselected large cohort of patients diagnosed with a neurodegenerative disease or knowing that the disease will develop in the future was directly asked about their advanced care planning and wishes surrounding the end of life. We conclude that many HD patients have end-of-life wishes and that we were not able to identify a certain group of patients to whom these questions should be specifically directed. Although HD is a neurodegenerative disease with cognitive decline, it is evident that many patients think about their future, the end of life and about ways to have a say in the end of life. Furthermore, because HD and other neurodegenerative diseases are relentless and result in difficulties with decision making in due course, we advise to discuss end-of-life wishes early in the disease and recommend that physicians initiate these conversations with every patient. The fact that less advanced patients, with higher education level more frequently refer to euthanasia or PAS shows that it is important to talk about these wishes from the earliest stages of HD but also of other neurodegenerative diseases when cognition is still intact and decisions can be made, based on the fact that as disease advances patients progressively loose insight. The finding that less than half discussed their wishes with a physician but many with family members shows that physicians should raise the awareness that the topic is of interest and of importance to the physician.

We recommend that these conversations should not only be held with patients in The Netherlands, where euthanasia and PAS are legal, but also in every other country, because we feel that nearly all patients, irrespective of the country where they reside, will have thoughts or wishes about the end of life. We expect that also patients in countries where euthanasia and PAS are illegal or under debate think about staying in control of their life and want to retain quality of life. For this reason we suggest physicians should ask every patient with HD or any other neurodegenerative disease early in their disease course about their fears, their wishes and thoughts for the future. When talking about euthanasia or PAS is not an option, a physician may discuss for example the desirability of tube feeding, or the use of antibiotics in the later stages of their disease. In the Netherlands physicians are designated to make medical decisions about starting or withholding treatment and following treatment directives and non-treatment directives, therefore physicians should hold these conversations. Law and practice in other countries determines which health care professional is the designated person to initiate these conversations.

Furthermore we suggest that wishes for the end of life other than euthanasia or PAS should be studied in patients with HD in order to examine which questions to ask patients in the early stages of the disease. We suggest that these studies should be carried out using qualitative techniques at first.

## Acknowledgements

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## REFERENCES

(1) [Termination of life on request and assisted suicide act.], 2002.

(2) van Tol DG, Rietjens JA, van der Heide A. Empathy and the application of the ‘unbearable suffering’ criterion in Dutch euthanasia practice. Health Policy 2012 May;105(2-3):296-302.

(3) van der Heide A, Onwuteaka-Philipsen BD, Rurup ML, Buiting HM, van Delden JJ, Hanssen-de Wolf JE, et al. End-of-life practices in the Netherlands under the Euthanasia Act. N Engl J Med 2007 May 10;356(19):1957-65.

(4) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(5) Borgsteede SD, Deliens L, Graafland-Riedstra C, Francke AL, van der Wal G, Willems DL. Communication about euthanasia in general practice: opinions and experiences of patients and their general practitioners. Patient Educ Couns 2007 May;66(2):156-61.

(6) de Boer ME, Hertogh CM, Droes RM, Jonker C, Eefsting JA. Advance directives in dementia: issues of validity and effectiveness. Int Psychogeriatr 2010 Mar;22(2):201-8.

(7) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. Advance directives for euthanasia in dementia: how do they affect resident care in Dutch nursing homes? Experiences of physicians and relatives. J Am Geriatr Soc 2011 Jun;59(6):989-96.

(8) Dees MK, Vernooij-Dassen MJ, Dekkers WJ, Elwyn G, Vissers KC, van WC. Perspectives of decision-making in requests for euthanasia: A qualitative research among patients, relatives and treating physicians in the Netherlands. Palliat Med 2013 Jan;27(1):27-37.

(9) A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington’s disease chromosomes. The Huntington’s Disease Collaborative Research Group. Cell 1993 Mar 26;72(6):971-83.

(10) Fisher ER, Hayden MR. Multisource ascertainment of Huntington disease in Canada: Prevalence and population at risk. Mov Disord 2013 Oct 1;n/a.

(11) www.cbs.nl, 2014.

(12) http://www.euthanasiecommissie.nl/contact/, 2014.

(13) Booij SJ, Engberts DP, Rodig V, Tibben A, Roos RA. A plea for end-of-life discussions with patients suffering from Huntington’s disease: the role of the physician. J Med Ethics 2013 Oct;39(10):621-4.

(14) Booij SJ, R+¦dig V, Engberts DP, Tibben A, Roos RAC. Euthanasia and Advance Directives in Huntington’s Disease: Qualitative Analysis of Interviews with Patients. Journal of Huntington’s Disease 2013 Jan 1;2(3):323-30.

(15) Britten N. Qualitative interviews in medical research. BMJ 1995 Jul 22;311(6999):251-3.

(16) Strauss AL, Corbin J. Basics of qualitative research: techniques and procedures for developing grounded theory. London: Sage Publications; 1998.

(17) Groenvold M, Petersen MA, Aaronson NK, Arraras JI, Blazeby JM, Bottomley A, et al. The development of the EORTC QLQ-C15-PAL: a shortened questionnaire for cancer patients in palliative care. Eur J Cancer 2006 Jan;42(1):55-64.

(18) Shoulson I, Fahn S. Huntington disease: clinical care and evaluation. Neurology 1979 Jan;29(1):1-3.

(19) Unified Huntington's Disease Rating Scale: reliability and consistency. Huntington Study Group. Mov Disord 1996 Mar;11(2):136-42.

(20) Folstein MF, Folstein SE, McHugh PR. "Mini-mental state". A practical method for grading the cognitive state of patients for the clinician. J Psychiatr Res 1975 Nov;12(3):189-98.

(21) van Delden JJ, van der Heide A, van de Vathorst S, Weyers H, van Tol DG. Kennis en opvattingen van publiek en professionals over medische besluitvorming en behandeling rond het einde van het leven Het KOPPEL-onderzoek. ZonMW, Den Haag; 2011.

(22) Rurup ML, Pasman HR, Onwuteaka-Philipsen BD. [Advance euthanasia directives in dementia rarely carried out. Qualitative study in physicians and patients]. Ned Tijdschr Geneeskd 2010;154:A1273.

(23) Dupuis HM. Professional autonomy: A stumbling block for good medical practice. An analysis and interpretation. Theor Med Bioeth 2000 Sep;21(5):493-502.

(24) Jochemsen H, Ten Have H. The autonomy of the health professional: An introduction. Theor Med Bioeth 2000 Sep;21(5):405-8.

(25) Gastmans C, De Lepeleire J. Living to the bitter end? A personalist approach to euthanasia in persons with severe dementia. Bioethics 2010 Feb;24(2):78-86.

(26) Hardwig J. Is there a duty to die? Hastings Cent Rep 1997 Mar;27(2):34-42.

(27) Rurup ML, Onwuteaka-Philipsen BD, van der Heide A, van der Wal G, Deeg DJ. Frequency and determinants of advance directives concerning end-of-life care in The Netherlands. Soc Sci Med 2006 Mar;62(6):1552-63.

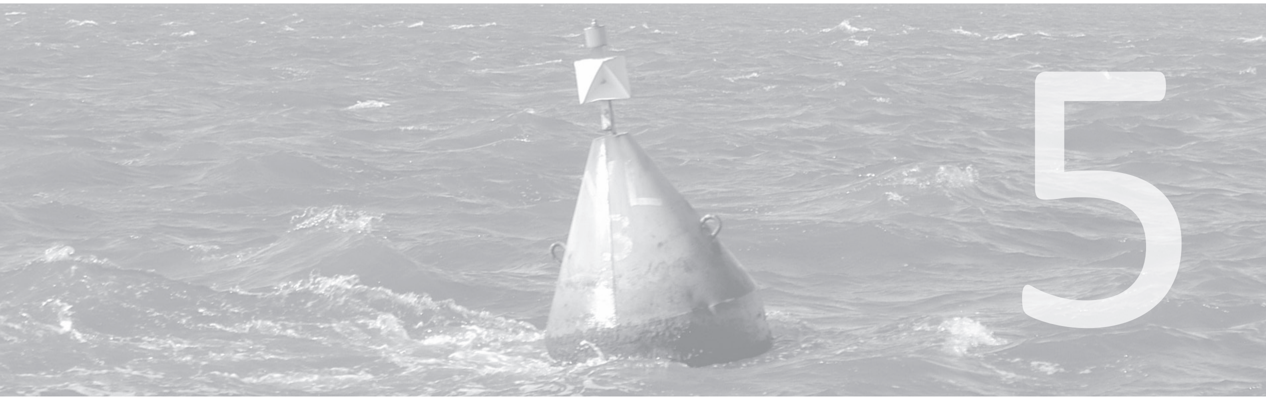
(28) Dellefield ME, Ferrini R. Promoting Excellence in End-of-Life Care: lessons learned from a cohort of nursing home residents with advanced Huntington disease. J Neurosci Nurs 2011 Aug;43(4):186-92.

(29) Aghababaei N, Wasserman JA. Attitude Toward Euthanasia Scale: Psychometric Properties and Relations With Religious Orientation, Personality, and Life Satisfaction. American Journal of Hospice and Palliative Medicine 2013 Jan 23.

(30) Leppert W, Gottwald L, Majkowicz M, Kazmierczak-Lukaszewicz S, Forycka M, Cialkowska-Rysz A, et al. A Comparison of Attitudes Toward Euthanasia Among Medical Students at Two Polish Universities. J Canc Educ 2012;1-8.

(31) Bulow HH, Sprung CL, Baras M, Carmel S, Svantesson M, Benbenishty J, et al. Are religion and religiosity important to end-of-life decisions and patient autonomy in the ICU? The Ethicatt study. Intensive Care Med 2012 Jul;38(7):1126-33.

(32) Becker J, Hart dJ. Godsdienstige veranderingen in Nederland. Verschuivingen in de binding met de kerken en de christelijke traditie. Den Haag: Sociaal en Cultureel Planbureau; 2006.



# A plea for end-of-life discussions with patients suffering from Huntington's disease: the role of the physician

Suzanne J Booij
Dick P Engberts
Verena Rödig
Aad Tibben
Raymund AC Roos

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## ABSTRACT

Euthanasia and physician-assisted suicide (PAS) by request and/or based on an advance directive are legal in The Netherlands under strict conditions, thus providing options to stay in control and choose ones end-of-life for patients with Huntington's disease (HD) and other neurodegenerative diseases. HD is an inherited progressive disease characterised by chorea and hypokinesia, psychiatric symptoms and dementia. From a qualitative study based on interviews with 15 physicians experienced in treating HD, several ethical issues emerged. Consideration of these aspects leads to a discussion about the professional role of a physician in relation to the personal autonomy of a patient. Such a discussion can raise awareness that talking about end-of-life wishes with an HD patient is part of the legal, professional and moral responsibility of the physician, and that a letter of intent on behalf of the physician can improve active participation in the process. Discussion of these issues can help to advance the debate on euthanasia and PAS in HD and other neurodegenerative diseases.

## INTRODUCTION

In The Netherlands the euthanasia act was approved by parliament in 2002. Euthanasia and physician-assisted suicide (PAS) are considered to be legitimate provided strict conditions are met. Approximately 7% of Dutch inhabitants have an advance directive, usually consisting of a euthanasia request, mainly as a precautionary measure in anticipation of possible dementia.[1] Euthanasia is legitimate based on an advance directive, but it is seldom performed, because of response shift or loss of competence and because physicians hesitate to perform euthanasia based solely on an advance directive.[1-4] Response shift means adapting to the disease or deviating from anticipatory beliefs.

Huntington's disease (HD) is an inherited autosomal dominant progressive neurodegenerative disease, characterised by chorea and hypokinesia, psychiatric symptoms and progressive dementia. Age of onset is usually between 30 and 50 years of age.[5] Despite symptomatic treatment, no cure is available and all patients will eventually become totally dependent for all daily life activities. There are approximately 1,700 HD patients in The Netherlands and about 5,000-8,000 persons are at risk. Between six and ten euthanasia requests in cases of HD have been granted in The Netherlands each year for the past five years.[6]

As HD is inherited, there is no doubt about the diagnosis after genetic testing and the course of the disease is usually known to patients and their families, HD can be used as an example for thinking about end-of-life questions in a disease where cognitive decline is a key symptom and the ethical dilemmas surrounding this subject. By exploring these questions with patients, especially in the premanifest phase, solutions can probably be implemented and may also enhance treatment and guidance in patients with other neurodegenerative diseases. Besides clinical experience we are not aware of studies on euthanasia or PAS or other end-of-life questions in patients with HD.

The aim of this study is to explore the role of the physician when it comes to talking about end-of-life wishes and the value of an advance directive, including its content. From the results we propose a new strategy for the treatment and guidance of patients and for dealing with end-of-life wishes in patients with other neurodegenerative diseases.

# METHODS

Between January 2011 and May 2011 a semi-structured interview with fifteen physicians was conducted. The physicians were recruited because of their experience with HD and their different fields of expertise. Our sample consisted of four general practitioners, two psychiatrists, two neurologists and seven nursing home physicians (NHP). Respondents were interviewed based on the method of purposive sampling, a commonly used form of non-probability sampling. Sample size relies on the concept of “saturation” or the point at which no new thematic content is observed in the data. Thematic saturation was reached after approximately twelve interviews. Although based on a topic list, the items covered by the interview could be expanded on during the interview phase, if new information or new insights emerged.[7] Interview topics included the communication about euthanasia/PAS with patients, experience with and moral considerations about euthanasia or PAS in HD, experience with advance directives in HD, management of symptoms of HD, in particular cognitive decline and psychiatric symptoms. We also addressed the participants’ perception of the reasons why patients want to talk about this subject with their physician and possible limitations to executing an advance directive. The questions were directed towards cases of HD, but sometimes other experiences were also mentioned and discussed. All interviews were recorded and fully transcribed. As our study was explorative, we used open coding. The transcripts were read and categorised into similar subject areas using inductive coding.[7] Representative quotations were chosen to demonstrate the themes identified.

# RESULTS

All physicians have more than 5 years experience in their field of expertise. Two GPs had a HD patient in their practice and performed euthanasia and two have a HD patient with whom they discuss their advance directive regularly. The nursing home physicians all work or worked in specialized HD units of a nursing home, except one. Three nursing home physicians performed euthanasia or PAS in HD. The psychiatrists and neurologists are specialized in HD (Table 5.1).

## Conversations about end-of-life wishes

Most respondents feel that talking about end-of-life wishes can bring peace of mind to a patient and that having the conversation sometimes makes the subject less pressing.

**Table 5.1** Characteristics of physicians

| Respondent (R)/ Type of physician | Number of patients with HD in the practice | Supportive of euthanasia/ Participation in euthanasia in HD | Raises the issue of advance directives | Raises the issue of euthanasia with patients |
|---|---|---|---|---|
| 1, GP | 1 | Yes/yes | Yes | Yes |
| 2, GP | 1 | Yes/ yes | Yes | Yes |
| 3, GP | 1 | Yes/ yes | Reacts to questions | Reacts to questions |
| 4, GP | 1 | Yes/ yes | Yes | Reacts |
| 5, NHP | >50 | Yes/ yes | Yes | yes |
| 6, NHP | >10 | Yes/ yes | Yes | yes |
| 7, NHP | >50 | Yes/ yes | Yes | yes |
| 8, NHP | >25 | Yes/ no | Yes | Reacts |
| 9, NHP | >70 | Yes/ yes | Yes | yes |
| 10, NHP | >100 | Yes/ yes | Yes | Yes |
| 11, NHP | >30 | Yes/ yes | No, reacts | No, reacts |
| 12, psychiatrist | >50 | Yes/ no | Reacts | Reacts |
| 13, psychiatrist | >50 | Yes/yes | No, reacts | Reacts |
| 14, neurologist | 1 | Yes/ yes | No, reacts | No, reacts |
| 15, neurologist | > 30 | Yes/no | No | No |

HD = Huntington’s Disease, GP = general practitioner, NHP = nursing home physician.

Respondent (R)13: “If subjects are not talked about, they become bigger. So I believe that if you discuss it regularly it can prevent euthanasia, or to put it differently, it can have the result that patients decide they no longer want euthanasia; it is no longer necessary.” R 4: “A more peaceful state of mind can be achieved when a patient talks about euthanasia. Peace in the knowledge that they will get help when needed. If counselling and support are provided in most cases you will not reach the point of really considering euthanasia.”

Some respondents indicated some hesitation to start the conversation. R 13: “I talk about the subject when a patient starts the conversation. I do not enter into the conversation spontaneously. Not because I think it is difficult, but because I leave it up to the patient to express themselves about this subject.” R 15: “I do not consider talking about euthanasia to be part of my job. Unless the conversation is initiated by the patient.” R 14: “I never ask about this myself. Because I feel that a patient should start the conversation. I thought about this for a long time, but I feel that if a patient is afraid to talk about the subject of euthanasia, then the patient is not ready for it.”

On the other hand, respondents remarked that euthanasia and/or PAS is an option a patient should know about. R 2: “I told my patient, I’m open to talk about the subject and to perform euthanasia, but it is a journey we should make together.” R 1: “I promised my patient I would help him, although this promise was emotionally difficult for me as a physician.”

### Advance directives and the role of the physician

The respondents explore the reasons for thinking about drawing up an advance directive with their patients. R 6: “The advantage of this disease is that patients do have some idea about what their future will be like. Many of them think about the end of life.”

The physicians usually also explain to the patient what they can expect from their physician, especially if the questions concern euthanasia or PAS. The physicians explain that drawing up an advance directive is letting the physician know what your wishes and values are, but it is not a guarantee that the result wished for by the patient will be reached. R 4:“He trusts us that we will help him in due course. Although I always tell all my patients that euthanasia is not a right.” General practitioners usually discuss the advance directive frequently when the wish becomes current or pressing and when it is brought up by the patient. Otherwise the advance directive becomes part of the patients’ file.

When the interviewer mentioned that euthanasia or PAS is possible based on a request by advance directive, several physicians responded that an advance directive was a useful and helpful document in determining the will of the patient, but not a document on which they would act. R 6: “It is the actual wish that counts. If this wish no longer exists, the advance directive also becomes invalid.” R 10: “I respond to the will expressed during conversation. But I have to say, if someone has taken the effort to draw up an advance directive, it means something. But the actual will is more important.”

## DISCUSSION

In The Netherlands, patients are allowed to make choices regarding end-of-life wishes and to express these to their physician. Physicians in this study recognise the fact that HD patients think about the end of life and have end-of-life wishes, but do not initiate a conversation about this subject. And although advance directives are used, physicians do not act solely on the basis of an advance directive.

We find there are three reasons why a physician should have a duty to explicitly discuss end-of-life wishes with patients diagnosed with HD.

1. From a legal point of view a physician is bound to inform the patient about his prospects with regard to the disease and treatment options (Civil Code art 7:448, paragraph 2). In this part of the Civil Code, parliament defined patient rights. Parliament also decided to place the physician at the centre of medical treatment. During the development of the euthanasia act however, parliament decided that euthanasia should be excluded from normal medical treatment. Parliament chose the position of the physician and his conflict of duties with respect to euthanasia (respect for human life versus the wish of a suffering patient) as the central issue and not the patient and his personal autonomy.[8] This choice by parliament also means that a physician is not obliged to talk about end-of-life wishes and try to find a way to help his patient with such a wish. A patient cannot claim or expect euthanasia from his physician; it is a request, without legal binding consequences.[8] But we argue that the legal obligation to inform the patient about his prospects can include the obligation to inform the patient about his prospects and options at the end of life.
2. From a professional point of view, Dutch guidelines state that a physician should act in the best interests of the patient, although sometimes the clinician's integrity can be in conflict with patient autonomy.[9;10] The Royal Dutch Medical Association (KNMG) wrote a paper on the professional role of the physician, advising physicians to initiate the conversation about end-of-life wishes at an early stage.[11] The KNMG stated that a physician should consider talking about this as part of his professional responsibility. From the present study one can conclude that the respondents are aware of the fact that it is important to talk about the patient's wishes, but that actually initiating the conversation remains difficult, despite KNMG guidelines.
3. We argue that it is not only a legal and professional responsibility but there is also a (moral) obligation to talk about end-of-life wishes.

HD patients ask questions about their prospects (including the end of life). They receive a professional response from their physician. Within a physician-patient relationship professionalism will be partly fed by moral considerations. And moral considerations will become dominant outwith the professional domain. In a physician-patient relationship a physician also is a fellow human being and not just a professional. Helping your patient and having a relationship, especially in the context of HD, means it is a matter of course to face the end of life. Informing the patient about his prospects can be extended to informing about the options at the end of life. Especially when respondents recognise that talking about end-of-life wishes can improve quality of life and that euthanasia or PAS are options a patient should be aware of. In our opinion talking about quality of life and retaining that quality until the end of life leads to talking about quality of dying. If the physician does not consider talking about end-of-life wishes or euthanasia as part of his job, he may have quite a strict opinion about a physician's professional task without taking moral considerations into consideration.

If a physician is unable to reconcile euthanasia with his conscience, a referral to another physician could be a way of taking the professional and moral responsibility and acting in the best interest of the patient. In a society where information concerning euthanasia is widely available we suggest that a physician, although not bound to reach the desired point but advised to refer when necessary, should consider it his job to talk about options at the end of life. Nevertheless, it is clear that a patient with such a request sometimes has difficulty finding a physician.[3;8;12;13] Thus even when the law only affirms the physician's obligation to inform about the prospects of the disease, and recognises the conflict of duties, we argue that a physician could have a moral obligation to re-evaluate his opinion about euthanasia or PAS and his response to patients asking for it.

Although legal and professional guidelines and regulations limit the role of other health care professionals in this area they can be very important for a patient. Caregivers, such as nurses, may be in closer contact with the patient than the physician. The team can fulfil an important role in signalling and interpreting the wishes of the patient and sometimes a team member is the first person to whom a patient expresses his wishes. Also other health care professionals may be the first to signal and communicate this to the physician.

## Advance directives

The conversation between patient and physician about end-of-life wishes can result in an advance directive. Especially in neurodegenerative disease such as HD, it is advisable to describe, determine and record the wishes, and the KNMG in fact advises physicians to enter into a conversation when a patient produces an advance directive and not just file the document.[11] The wishes can be documented by the patient himself or by the physician after a conversation and should be attached to the medical record. From the response concerning the applicability of advance directives it is possible to suggest that some physicians seem to lack knowledge, especially regarding the applicability of advance directives and euthanasia in dementia. Some physicians see the advance directive only as a helpful document, as also reported in earlier research.[1;3;12] Although the law provides a possibility to act, based solely on an advance directive, several reasons have been identified in literature why physicians seldom do. Reasons are for example a difficulty determining the unbearableness of the suffering and the absence of a (present) wish of the person involved. We add a possible lack of knowledge to these reasons. Not only a lack of knowledge on the possibility provided for in the law, but on how to fulfil the six requirements of the law on euthanasia and replace the request by an advance directive, and how to handle cognitive decline, response shift and psychiatric symptoms.

## Letter of intent on behalf of the physician

In order to enhance involvement and awareness of the possibilities provided for within the law, we propose a letter of intent on behalf of the physician. A suggestion to or obligation for the physician to draw up a document himself might result in a more active participation of the physician. General objections have been discussed and overcome and the physician is committed to make an effort to follow the patient's wishes. The physician is still able to return to this subject, but only on the basis of specific circumstances and not general objections.

This incentive could be stated in a letter of intent for the physician; a letter in which the patient and the physician agree upon a commitment of best intents (Table 5.2). Of course there is never a duty to achieve a given result, but if a commitment of best intents is written down and agreed upon it is impossible for a physician to ignore the subject at a later stage. This is also the moment when the physician can explore the morals, values

**Table 5.2** Suggested content of the letter of intent

| | |
|---|---|
| 1 | Date of conversation: documentation of content of conversation |
| 2 | Statements about the fact that the physician will feel obliged and committed to this subject or refer the patient to a colleague if necessary. |
| 3 | Education of the patient by talking through the legal requirements |
| 4 | If and which persons will be involved when talking through the wishes |
| 5 | Exploration of values and wishes of the patient: which aspects of quality of life are crucial for the patient |
| 6 | Arrangements on recurrence (frequency) of this conversation and renewal of the advance directive |

and wishes of the patient and when the patient can be educated on the subject, about the restrictions and requirements of the law, about what to expect from his physician and how to keep the advance directive up to date. Physician and patient can have a conversation about response shift and how to handle it. They can make arrangements on how often they will discuss the advance directive.

When exploring the wishes and values of the patient, a physician will also talk about the subject of suffering, one of the requirements of the law (Penal Code art. 293 and Law on Review of Euthanasia and PAS).[8] The concept of suffering is divided into two elements: unbearable suffering, which is a subjective opinion, and which must be understandable 'objectively' for a physician and a lack of prospect of improvement.[8] The lack of prospect of improvement is defined as the absence of acceptable treatment possibilities, the disease cannot be cured and symptoms cannot be relieved.[8]

The euthanasia law in the Netherlands is based (morally) on the principle of beneficence and (in terms of criminal law) the necessity defence. When a patient doesn't ask for euthanasia, euthanasia is not allowed and subject to a murder charge. Physician integrity combined with the requirements of the law prevents euthanasia in cases of 'tired of living'.[8] When we look at HD, where all criteria for euthanasia can be met, it is the physician who eventually has to decide if the suffering is unbearable to such an extent that euthanasia is justified. The recurrence of conversation about the wishes of the patient, including his view on quality of life can take the physician along the way towards a desired end of life. The physician can signal a possible response shift. The professional integrity of the physician partly determines the scope of applicability of end-of-life wishes.[14] By discussing and describing what is acceptable, the physician's his frame of mind can partly be influenced, without the loss of professional integrity.

## CONCLUSION

Euthanasia and PAS are still matters subject to extensive discussion in The Netherlands. According to the physicians interviewed, euthanasia is an option patients should have when suffering from a neurodegenerative disease such as HD. Despite the fact that the respondents provide arguments for initiating a conversation about the options a patient has, most physicians do not take this step. In this article we suggest that there can be a legal, professional and moral obligation for a physician to talk about the subject, especially in the case of neurodegenerative diseases such as HD. The physician should be aware of the fact that although euthanasia is not a normal medical treatment, it is part of his obligation to educate the patient about his disease and his prospects and it may be his responsibility to educate the patient about the possibilities of choosing the end of life. This obligation can be seen as a commitment of best intents, laid down in a letter of intent by the physician.

We suggest further research to be carried out into the wishes of patients, not only to improve treatment, but also to explore the expectations they have of their physician. We also suggest physicians be better educated about end-of-life wishes and the possibilities provided for in the law, thus assisting them in improving the way they help their patients to deal and cope with a devastating disease such as HD as well as other neurodegenerative diseases.

## REFERENCES

(1) Rurup ML, Pasman HR, Onwuteaka-Philipsen BD. [Advance euthanasia directives in dementia rarely carried out. Qualitative study in physicians and patients]. Ned Tijdschr Geneeskd 2010;154:A1273.

(2) Hertogh CM, de Boer ME, Droes RM, Eefsting JA. Would we rather lose our life than lose our self? Lessons from the Dutch debate on euthanasia for patients with dementia. Am J Bioeth 2007 Apr;7(4):48-56.

(3) Rurup ML, Onwuteaka-Philipsen BD, van der HA, van der WG, van der Maas PJ. Physicians' experiences with demented patients with advance euthanasia directives in the Netherlands. J Am Geriatr Soc 2005 Jul;53(7):1138-44.

(4) van Delden JJ. The unfeasibility of requests for euthanasia in advance directives. J Med Ethics 2004 Oct;30(5):447-51.

(5) Bird TD. Outrageous fortune: the risk of suicide in genetic testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1289-92.

(6) http://www.euthanasiecommissie.nl/contact/. 2014.

(7) Britten N. Qualitative interviews in medical research. BMJ 1995 Jul 22;311(6999):251-3.

(8) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(9) Lantos J, Matlock AM, Wendler D. Clinician integrity and limits to patient autonomy. JAMA 2011 Feb 2;305(5):495-9.

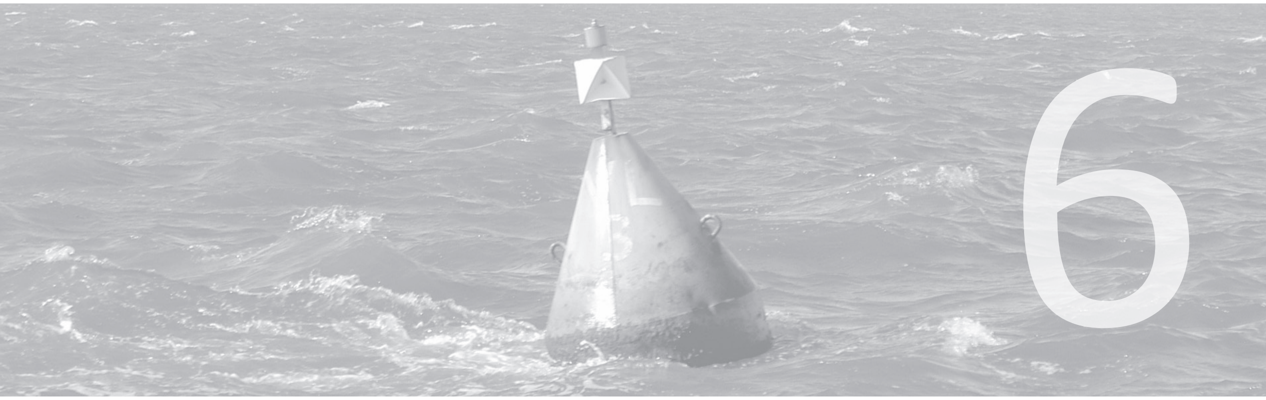
(10) Wendler D. Are physicians obligated always to act in the patient's best interests? J Med Ethics 2010 Feb;36(2):66-70.

(11) [Early conversations about end-of-life wishes]; http://knmg.artsennet.nl/Diensten/knmgpublicaties/KNMGpublicatie/Handreiking-Tijdig-praten-over-het-overlijden-2011.htm. 2011.

(12) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. Advance directives for euthanasia in dementia: how do they affect resident care in Dutch nursing homes? Experiences of physicians and relatives. J Am Geriatr Soc 2011 Jun;59(6):989-96.

(13) Pasman HR, Rurup ML, Willems DL, Onwuteaka-Philipsen BD. Concept of unbearable suffering in context of ungranted requests for euthanasia: qualitative interviews with patients and physicians. BMJ 2009;339:b4362.

(14) Billings JA, Krakauer EL. On Patient Autonomy and Physician Responsibility in End-of-Life Care. Arch Intern Med 2011 May 9;171(9):849-53.



# Advance Care Planning and decision making for the end of life in Huntington's disease: dilemmas for doctors

Suzanne J Booij
Dick P Engberts
Aad Tibben
Raymund AC Roos

Submitted.

# ABSTRACT

**Objective:** To obtain in-depth information about problems physicians encounter when talking about advance care planning (ACP), in particular end-of-life wishes, euthanasia or physician assisted suicide (PAS), in patients with a neurodegenerative disease such as Huntington's Disease (HD).

**Design and participants:** Semi-structured interviews in 2011 with a purposive sample of 15 physicians familiar with HD; general practitioners, nursing home physicians and medical specialists.

**Results:** According to the physicians, HD-patients increasingly show an interest in discussing end-of-life questions. We identified three patterns of how physicians evaluate decision making about the end of life. These patterns are based on different views on dealing with cognitive decline, psychiatric symptoms and signs and response shift. The evaluation and handling of these symptoms and signs in light of ACP proved to be specialty-specific and based upon a good and longstanding physician-patient relationship. 'Admission to a nursing home' as unbearable suffering elicits a different response from physicians in different specialties.

**Conclusions:** Loss of decision making capacities, psychiatric symptoms and signs and response shift are themes that apply to all physicians who are engaged in ACP in neurodegenerative diseases. The difficulties signaled and detected are not only applicable to physicians in the Netherlands, but extent to physicians in other countries as well when decisions for the end of life must be made. A firm and possibly longstanding physician-patient relationship with knowledge of a patient's past, present, expected future and values and goals can be of importance for all physicians in decision making.

# INTRODUCTION

Huntington's disease (HD) is a progressive neurodegenerative disease, caused by an autosomal dominant inherited expansion of a CAG-repeat on chromosome 4. The disease is characterized by chorea and hypokinesia, psychiatric symptoms and progressive cognitive decline leading to dementia.[1;2] There is no cure for this devastating disease and all patients eventually will become totally dependent for all daily life activities. Pneumonia is the primary cause of death, suicide is the secondary.[1;3;4] The rate of suicide is 5-12 times higher compared to the general population.[3-5] The prevalence of HD in the Netherlands is approximately 1,700. The estimated amount of persons at 50% risk is 5,000-8,000.

In the Netherlands euthanasia or physician assisted suicide (PAS) is legal under strict conditions after the euthanasia act was approved by parliament in 2002.[6] Euthanasia and PAS are defined as death brought upon by a physician, only after a direct request from the patient. Parliament decided that euthanasia is not a patient's right, but a request and a physician's competence. The physician decides to either or not respond to the patient's request.[7] The first condition of the law on the legalization of euthanasia and PAS is that the request must be voluntary and well-considered. If a physician has doubts about these requirements, he is supposed to make extra inquiries and for example ask for a psychiatric consultation. This applies especially when there are problems with cognition, communicative impairments or a psychiatric disorder. The second requirement of due care is the requirement of unbearable suffering with no prospect of improvement. Unbearable suffering is 'subjective', it is the patient's perspective that defines if suffering is unbearable. Whether the suffering is without prospect of improvement is also part of the medical expertise of the physician.[7] Earlier research showed that patients put more emphasis on psychosocial factors of suffering, whereas physicians refer more to physical suffering.[8] A difficult element regarding the element of unbearable suffering is the fact that it is subjective for the patient, but it must be 'recognizable' for the physician to some extent.[9;10] The other requirements are extensively described in literature.[6;7]

Between 6 and 10 requests for euthanasia or PAS in case of HD were granted in the Netherlands each year in the past five years.[6] Besides clinical experience there are no studies on euthanasia or PAS or other end-of-life questions in patients with HD.

With an increasing prevalence of neurodegenerative diseases, and usually limited treatment options, possibilities to have a say in ones end of life, for example euthanasia or PAS could enhance patient's quality of life and of dying.

The first aim of this study is to explore which problems physicians encounter when talking about end-of-life wishes, euthanasia or PAS with their HD patients. The second aim is to explore how physicians meet the wish of their patient, given that HD is a neurodegenerative disease characterized by cognitive decline and psychiatric symptoms.

## METHODS

The study consisted of semi-structured interviews with fifteen physicians who are experienced with the treatment of HD patients; four general practitioners (GP), two neurologists, two psychiatrists and seven nursing home physicians (NHP). Physicians were selected because of their knowledge of HD and were contacted by email. None of the contacted physicians declined. Interviews were conducted in 2011 and took place in their practice. 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.[11-13] Physicians were interviewed based on the method of purposive sampling, the most commonly used form of nonprobability sampling.[14] Sample size relies on the concept of "saturation" or the point at which no new information or themes are observed in the data. Saturation was reached after the 12th interview. All interviews were performed by the same interviewer and took approximately one hour to complete. The interviews consisted of questions concerning the presence of end-of-life wishes, the conversations between patient and physician about the end of life, why such questions emerged and the problems experienced with this subject, such as cognitive decline, psychiatric symptoms and response shift, i.e. adapting to the disease or deviating from anticipatory beliefs. The interviews lasted approximately one hour.

All interviews were recorded, fully transcribed and subjected to qualitative analysis. As our study was explorative, we used open coding to identify relevant concepts. A second independent researcher read and coded all the interviews. The list of codes was discussed between the first author and the independent researcher. Examples of codes are cognitive changes, response shift, advance directives and reasons for thoughts about euthanasia by the patients and reactions from the physicians when these questions are asked.

# RESULTS

All physicians had at least five years of experience in their field of expertise. Two GPs had a HD patient in their practice and performed euthanasia and two have a HD patient with whom they discuss the patient's advance directive regularly. All but one of the nursing home physicians work or worked in specialized HD units of a nursing home. Three nursing home physicians had experience with performing euthanasia or PAS in HD. The psychiatrists and neurologists are specialized in HD (Table 6.1).

## Conversations about end-of-life wishes

In general some of the respondents have noticed that questions regarding end-of-life wishes have been increasingly raised by patients in the last couple of years (Table 6.2, Quotation 1). One physician mentioned that end-of-life wishes have always been present in HD patients, but are more frequently discussed after the euthanasia law was passed. Respondents recognized two groups of patients. One group of HD patients does not think or speak about end-of-life wishes; the other is well informed on the subject and has thought about it before talking to a physician (Table 6.2, quotation 2-4).

**Table 6.1** Physician characteristics

| Physician/Respondent | Specialty | Gender |
|---|---|---|
| 1 | GP | Male |
| 2 | GP | Male |
| 3 | GP | Female |
| 4 | GP | Female |
| 5 | Psychiatrist | Male |
| 6 | Psychiatrist | Female |
| 7 | Neurologist | Male |
| 8 | Neurologist | Male |
| 9 | NHP/GP | Male |
| 10 | NHP | Female |
| 11 | NHP | Female |
| 12 | NHP | Male |
| 13 | NHP | Female |
| 14 | NHP | Male |
| 15 | NHP | Female |

HD = Huntington's Disease, GP = General Practitioner, NHP = Nursing Home Physician.

**Table 6.2** Conversations about end-of-life wishes

| | |
|---|---|
| 1 | R10: "With our new out-patient clinic, I think questions will be raised increasingly. Patients approach us: "I want to make amends for the future." |
| 2 | R12: "Patients, who are able to think about the subject and have thought about the subject, start the conversation themselves." |
| 3 | R7: "It is only a small percentage of patients that ask questions about the end of life." |
| 4 | R11: "I notice that HD patients can be divided into two groups. The biggest group does not have a lot of understanding of the disease and disease awareness. And there are some people that are extremely occupied with this subject (red. Wishes for the end of life) and from those people I get a lot of questions or inquiries about euthanasia." |

## How to recognize the patient's wish in case of a neurodegenerative disease?

The physicians could be divided into three groups based on the responses regarding symptoms and signs of HD. The group of general practitioners represents the first group. When asked about cognitive changes and psychiatric symptoms and signs they indicated that they know their patient well and were able to determine if their patient was competent to make decisions. But they also mentioned that it is important to recognize and appreciate response shift over time (Table 6.3, quotation 1 and 2).

The second group of specialists (psychiatrists and neurologists) mentioned that when speaking with a HD patient it could be difficult to find out whether a patient is fully competent to express end-of-life wishes or whether the wishes are part of a psychiatric symptom such as depression or an obsessive thought (Table 6.3, quotation 3-5). Consequently, the value of the wish was questioned.

**Table 6.3** Determine the patient's wish: general practitioners and medical specialists

| | |
|---|---|
| 1 | R3: "She had always been clear, in every moment of choice and during the whole process of thinking about her wish." |
| 2 | R4: "To clarify his wishes, that has been a long process and he is more aware of his wishes now. And he has been consistent in his wishes for such a long period of time, that I would be surprised if that would change. But we also talked about a possible change, to let him know he can change his wishes any moment." |
| 3 | R5: "HD patients have difficulties to understand complex situations. A decision to opt for euthanasia and the process and procedures surrounding this choice are more difficult to understand for HD patients, in the stage when cognitive decline becomes clear. This is already possible early on in the disease." |
| 4 | R7: "Compulsive behavior is also part of HD. The question is if the request for euthanasia really is well considered or if it is part of an impulse of the compulsive behavior." |
| 5 | R8: "If a patient, with a partly disturbed perception, in light of his disease wants to make a decision with grave consequences, then the question remains if a physician should intervene." |

The third pattern was recognized within the group of nursing home physicians, who struggled the most with the value of the wish, competence and also with the value of the advance directives. They noticed that at the time of admittance the decision making capacity and competence had already diminished. Sometimes the admittance itself reflected a patients' response shift (Table 6.4, quotation 1-4). In contrast to the GPs and specialists, NHPs first see patients in the latter stages of the disease. In general, advance directives are presented to them, sometimes by family members (Table 6.4, quotation 5).

Neurologists, psychiatrists and NHPs agree with the option of euthanasia in HD, but consider the eventual performance as a task of the GP (Table 6.4, Quotation 6-7). However they acknowledged that a patient who expresses the need to talk about euthanasia or PAS shows some level of competence, which need should be appreciated and responded to accordingly.

## Suffering

According to the law, suffering should be unbearable to warrant euthanasia or PAS.

Most of the GPs could appreciate the wish of a patient for euthanasia in case of an unacceptable but inevitable move to a nursing home. Admission to a nursing home means for many patients full awareness of having reached the last stage of the disease

**Table 6.4** Determine the patient's wish: Nursing home physicians

| | |
|---|---|
| 1 | R15: "If there is cognitive deterioration, there is a shift in the judgment upon what is acceptable. With a lot of people, things are never as bad as they seem. In a nursing home euthanasia is not often a subject. Patients changed and shifted so much already." |
| 2 | R15: "Euthanasia is seldom performed. The main problem of the disease is the inability to make a plan. Important symptoms of HD are impulsiveness and behavioral disturbances and these symptoms can have a major impact on decision making capacities and competence." |
| 3 | R13: "When a patient is admitted here, their cognition deteriorated to such an extent that it is almost impossible to find out what the wishes were. Or the cognition is intact, but there are so many psychiatric symptoms that that prohibits a clear request for euthanasia." |
| 4 | R14: "Adaptation happens. You adapt to the situation. I can imagine that when it becomes impossible to stay at home, this is a next step of adaptation." |
| 5 | R10: "Some have made an advance directive, earlier, with the GP, they show it to me….It is difficult, what is being competent, with this strange disease." |
| 6 | R8: "I understand that patients would consider euthanasia. But I think euthanasia is something that belongs with the general practitioners." |
| 7 | R6: "I think that a GP or NHP knows a patient better, knows the circumstances, so it is easier to concur with the situation of a patient." |

with total dependency on others and no prospect of future improvement of the situation. Therefore, moving to a nursing home could qualify as unbearable suffering (Table 6.5, quotation 1 and 2).

On the other hand all but one of the NHPs have the experience that the transfer of an HD patient from his home to a nursing home could ease some of the problems they encountered at home and result in a lessening of suffering. Medical specialist had more diverse opinions, both being able to qualify the admission as suffering or as an option to ease suffering (Table 6.5, quotation 3-5).

**Table 6.5** Suffering, a different perspective

| | |
|---|---|
| 1 | R3: "Patients prefer to stay at home. And we try to make this possible with all kinds of care at home." |
| 2 | R12: "I can imagine that a nursing home for some patients is unacceptable." |
| 3 | R14: "As a GP you witness the disease process, in a nursing home we see the patient in an advanced phase. I believe that suffering is more visible for a GP and easier to understand." |
| 4 | R8: "As a physician you have to understand what marks the end of adaptation for a certain patient. And for a general practitioner that can be easier than for a NHP. A NHP sees patients who are very ill and the reference for a GP is the fairly healthy human or patient." |
| 5 | R9: "With a lot of people, things are never as bad as they seem. In a nursing home euthanasia is not a subject which is frequently brought up. Patients have changed and shifted so much already." |

## DISCUSSION

Physicians have observed that HD patients increasingly consider and accept euthanasia and PAS as an end-of life-option. Physicians are more open to respond to these considerations although they do not initiate with the exception of NHPs. All physicians distinguished between individuals who face their future disease prospects and wish to exert control over their lives, and those who avoid confrontation with the disease and live from day to day. GPs and medical specialists usually do not initiate a conversation about end-of-life wishes, contrary to NHPs who usually initiate conversations early after admission, with both the close relatives and the patients, where it is part of the procedure for advanced care planning (ACP).[15;16]

### Cognitive decline

When talking about ACP and wishes for the end of life, according to the law, a physician has to be certain that the request for euthanasia or PAS was voluntarily and well-considered.[17] To evaluate if a request was voluntary and well-considered in case of a neurodegenerative disease such as HD, a physician has to take cognitive decline and psychiatric symptoms into account. Especially in neurologic diseases, such as HD, cognitive deficits, communication deficits and psychiatric symptoms may impair the ability of patients to decide upon wishes for the end of life, euthanasia or PAS.[18] Nevertheless patients want to make decisions for themselves regarding health care and treatment for as long as they can.[19] Two main arguments are posed when considering ACP undertaken by a patient who loses capacities: 1) the patient who loses capacity is not the same person as the person who had capacities and 2) the patient with cognitive deficits still has autonomy and should not be held to previously expressed wishes.[20-24] Besides, a change in previously expressed end-of-life wishes may also be explained by response shift.

Patients suffering from dementia will gradually progress and become less competent and lose decision making capacities as the disease progresses.[25] On the other hand, other research showed that patients in the early stages of Alzheimer’s Disease are aware of their failing memory and are able to tell about their experiences with the disease.[22] Nevertheless the gradual progression allows the patient to adapt and adjust to the changing situation. This adaptation explains the second argument that because the patient’s personality changes, patients with cognitive deficits will not act in accordance with their earlier values and beliefs and thus should not be held to previously expressed wishes.[22;26] This adaptation is part of what the process of response shift beholds. Recognizing response shift is important when talking to patients and about their wishes for the end of life and making a decision with the patient. The GPs in this study reported that they were able to recognize cognitive changes and response shift. The GPs recognized the phenomenon of response shift but also mentioned they could adequately respond to this, just because they know their patient well and usually have treated and supported their patient for many years. If a long physician-patient relationship preceded the decision about euthanasia or PAS, a GP will be able to tell if the decision was well-considered and consistent.

In contrast to GPs, other physicians in this study got involved with the care and treatment of the patient after the diagnosis had been made and guidance and treatment became necessary. They had a different physician-patient relationship (specialists) or were only involved in the last stages of the disease (NPHs). Knowing if the wish was well-considered and consistent was often difficult to find out and knowing if response shift had occurred and the patient deviated from anticipatory beliefs proved difficult to determine, because of the missing longstanding physician-patient relationship. In case of HD these physicians mention cognitive decline and psychiatric symptoms and signs as difficult aspects to evaluate and to cope with when having conversations about ACP, euthanasia and PAS. Consequently, it was difficult to know if the present situation of the patient is compatible with a patient's values and goals.[27] Secondly, it was difficult to know if the present situation is the one for example previously talked about in the context of ACP or described in an advance directive, as was also described in a study about the sometimes problematic differences of views between physicians and family members/surrogates when making a decision for patients who have lost decision-making capacity.[28] Other research also found that in patients with dementia the lack of key moments to initiate conversations about ACP, the patient's lack of awareness of their diagnosis and prognosis and the fact that patients did not often initiate such discussions themselves were barriers to engage in ACP as a physician.[29]

## Psychiatric symptoms and signs

Psychiatric symptoms and signs were also mentioned as difficult aspects to cope with for specialists and NHPs when having conversations about euthanasia or PAS. HD is characterized by a broad range of psychiatric symptoms and signs including depression and suicidality.[30] Research showed that particular appreciation of information is impaired in depression.[31;32] Secondly there is a claim that talking about euthanasia or PAS with a patient suffering from psychiatric signs and symptoms reinforces loss of hope and demoralization. The argument states that by talking about euthanasia, the central element in the physician-patient relationship is removed, namely the hope of improvement by treatment.[33] In general, intervention to prevent suicide is indicated, because the wish for suicide can be part of a treatable sign. But sometimes a wish for suicide, and thus euthanasia or PAS can be reasonable. If the wish arises as a symptom of the disease itself, it is generally accepted that the disease should be treated. But if the wish is based on a rational evaluation of a patient's past, present and future, a request can be reasonable.[33] In HD most patients know about the development of the disease and their future prospects. Psychiatric symptoms and signs are part of the disease, but the wish for euthanasia or PAS can well result from their knowledge of the disease instead of their current psychiatric symptoms. From the answers of the NHPs we can deduct that distinguishing between signs of the disease and a rational evaluation of a patient's actual state proves to be difficult. Again a longstanding physician-patient relationship seems to be an important factor in dealing with psychiatric symptoms and signs and incorporating euthanasia and PAS in ACP. Guiding a patient for many years provides the opportunity to actually know the patient, his values and goals, to detect cognitive decline and psychiatric symptoms and signs and to discuss the consequences, relating to ACP and euthanasia and PAS, openly with the patient.

## Suffering

The specialists in our study could appreciate that a nursing home represents suffering and subsequently the wish for euthanasia. On the other hand several NHPs regard the admittance to a nursing home as a possible way of improvement of quality of life and thus as diminishing of suffering. Earlier research stated that working in a nursing home means that the perception of a life that is unacceptable or meaningful can change.[34] This contrast can result in a different evaluation of a euthanasia request. The admittance to a nursing home may in itself also represent response shift and thus acceptance instead of suffering, which can be investigated by evaluating an advance directive, if present, and reports on the patient’s recent responses to the disease process. The GPs or specialist’s decision to qualify suffering as unbearable may be based on empathizing with the patient’s anticipated thoughts and feelings regarding admission in a nursing home.[6] One may raise the issue that when distinguishing between these groups of physicians, part of the distinction is due to the fact the physician possibly agrees or sympathizes with the patient. That as a fellow human admittance to a nursing home would be undesirable and thus represent suffering.[33] This might be different sometimes for NHPs. To try to avoid too much of a subjective judgement, the law provided several guarantees for these circumstances. First it is required that an independent second physician gets in contact with the patient and evaluates all the requirements of due care. Secondly the suffering must be unbearable to this patient. The patient’s history, past, values and goals determine if suffering is unbearable. Therefore, although a physician must recognize and emphasize with the suffering to some extent, physicians should realize that work environment can change their views, but not always the views of the patient. Suffering can be referred to as ‘increasing physical deterioration and dependency’ and ‘loss of dignity’.[7;35] Moving to a nursing home can be the realization or materialization of ‘loss of dignity’ and ‘total dependency’, thus of suffering, for some. Moving to a nursing home can be a practical solution for a need, without offering a real improvement of quality of life, thus without changing the views of the patient.

A limitation of this study is the relatively small number of physicians interviewed which were selected in advance. On the other hand, this is more of a problem for quantitative than for qualitative research, as the use of purposive sampling allowed us to possibly detect all relevant themes without looking for a representative sample of the profession as a whole.

Advance care planning, including euthanasia and PAS can be difficult topics for physicians to initiate, evaluate and talk about with patients. A second limitation could be that we explored opinions of physicians about these topics, what they experience and what they say about the topic, instead of what they actually do. But talking about these topics revealed the problems they encounter, but moreover the problems they suspect or think are present. By exploring and recognizing these problems, we are be able to signal the issues and propose solutions for physicians to overcome these issues and to possibly diminish the struggle with topics such as cognitive decline, response shift or the judgment upon suffering.

We argue that the loss of decision making capacities, psychiatric symptoms and signs and response shift are themes that apply to all physicians who are engaged in advance care planning in patients with neurodegenerative diseases. Secondly we argue that the difficulties signaled and detected are not only applicable to physicians in the Netherlands, but extent to physicians in other countries as well when dealing with patients that have a disease where decisions for the end of life must be made. A firm and possibly longstanding physician-patient relationship with knowledge of a patient's past, present, future and values and goals can be of importance for all physicians in decision making.

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## REFERENCES

(1) Bird TD. Outrageous fortune: the risk of suicide in genetic testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1289-92.

(2) Unified Huntington’s Disease Rating Scale: reliability and consistency. Huntington Study Group. Mov Disord 1996 Mar;11(2):136-42.

(3) Almqvist EW, Bloch M, Brinkman R, et al. A worldwide assessment of the frequency of suicide, suicide attempts, or psychiatric hospitalization after predictive testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1293-304.

(4) Paulsen JS, Hoth KF, Nehl C, et al. Critical periods of suicide risk in Huntington’s disease. Am J Psychiatry 2005 Apr;162(4):725-31.

(5) Farrer LA. Suicide and attempted suicide in Huntington disease: implications for preclinical testing of persons at risk. Am J Med Genet 1986 Jun;24(2):305-11.

(6) http://www.euthanasiecommissie.nl. 2014. last accessed May 2014.

(7) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(8) Pasman HR, Rurup ML, Willems DL, et al. Concept of unbearable suffering in context of ungranted requests for euthanasia: qualitative interviews with patients and physicians. BMJ 2009;339:b4362.

(9) Pans E. De normatieve grondslagen van het Nederlandse euthanasierecht. Nijmegen: Wolf Legal Publishers; 2006.

(10) Youngner SJ, Kimsma GK, et al. Physician-Assisted Death in Perspective. assessing the Dutch Experience. New York: Cambridge University Press; 2012.

(11) Britten N. Qualitative interviews in medical research. BMJ 1995 Jul 22;311(6999):251-3.

(12) Strauss AL, Corbin J. Basics of qualitative research: techniques and procedures for developing grounded theory. London: Sage Publications; 1998.

(13) Malterud K. Qualitative research: standards, challenges, and guidelines. Lancet 2001 Aug 11;358(9280):483-8.

(14) Staines GL. The causal generalization paradox: The case of treatment outcome research. Review of General Psychology 2008 Sep;12(3):236-52.

(15) Borgsteede SD, Deliens L, Graafland-Riedstra C, et al. Communication about euthanasia in general practice: opinions and experiences of patients and their general practitioners. Patient Educ Couns 2007 May;66(2):156-61.

(16) Back AL, Starks H, Hsu C, et al. Clinician-patient interactions about requests for physician-assisted suicide: a patient and family view. Arch Intern Med 2002 Jun 10;162(11):1257-65.

(17) [Termination of life on request and assisted suicide act.] (2002).

(18) Sizoo EM, Grisold W, Taphoorn MJ. Neurologic aspects of palliative care: the end of life setting. Handb Clin Neurol 2014;121:1219-25.

(19) Hamann J, Bronner K, Margull J, et al. Patient participation in medical and social decisions in Alzheimer's disease. J Am Geriatr Soc 2011 Nov;59(11):2045-52.

(20) McGee A. Advance care planning for dementia patients. BMJ Support Palliat Care 2013 Jun;3(2):237-8.

(21) Davis JK. The concept of precedent autonomy. Bioethics 2002 Apr;16(2):114-33.

(22) de Boer ME, Droes RM, Jonker C, et al. [The lived-experiences of early-stage dementia and the feared suffering: an explorative survey]. Tijdschr Gerontol Geriatr 2010 Oct;41(5):194-203.

(23) DeGrazia D. Advance directives, dementia, and 'the someone else problem'. Bioethics 1999 Oct;13(5):373-91.

(24) Gastmans C, De Lepeleire J. Living to the bitter end? A personalist approach to euthanasia in persons with severe dementia. Bioethics 2010 Feb;24(2):78-86.

(25) Gather J, Vollmann J. Physician-assisted suicide of patients with dementia. A medical ethical analysis with a special focus on patient autonomy. Int J Law Psychiatry 2013 Sep;36(5-6):444-53.

(26) de Boer ME, Hertogh CM, Droes RM, et al. Suffering from dementia - the patient's perspective: a review of the literature. Int Psychogeriatr 2007 Dec;19(6):1021-39.

(27) Billings JA, Krakauer EL. On Patient Autonomy and Physician Responsibility in End-of-Life Care. Arch Intern Med 2011 May 9;171(9):849-53.

(28) Smith AK, Lo B, Sudore R. When previously expressed wishes conflict with best interests. JAMA Intern Med 2013 Jul 8;173(13):1241-5.

(29) De Vleminck A, Pardon K, Beernaert K, et al. Barriers to advance care planning in cancer, heart failure and dementia patients: a focus group study on general practitioners' views and experiences. PLoS One 2014 Jan 21;9(1):e84905.

(30) Hubers AA, Reedeker N, Giltay EJ, et al. Suicidality in Huntington's disease. J Affect Disord 2012 Feb;136(3):550-7.

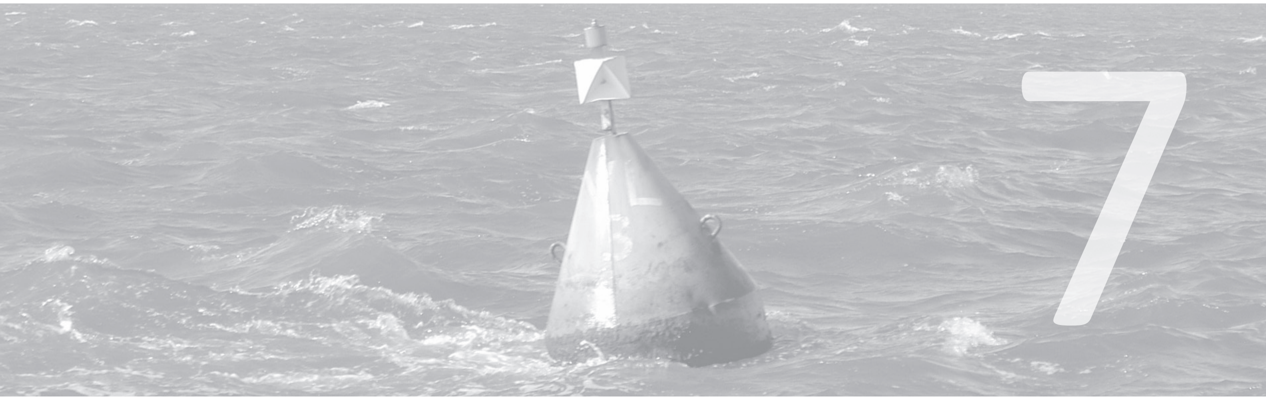
(31) Hindmarch T, Hotopf M, Owen GS. Depression and decision-making capacity for treatment or research: a systematic review. BMC Med Ethics 2013;14:54.

(32) Owen GS, Szmukler G, Richardson G, et al. Decision-making capacity for treatment in psychiatric and medical in-patients: cross-sectional, comparative study. Br J Psychiatry 2013 Dec;203(6):461-7.

(33) Berghmans R, Widdershoven G, Widdershoven-Heerding I. Physician-assisted suicide in psychiatry and loss of hope. Int J Law Psychiatry 2013 Sep;36(5-6):436-43.

(34) Touwen D. Former wishes and current desires. Demented patients and their family members’ effort to decide what they would have wanted. In: Schermer M., Pinxten W., eds. Ethics, Health Policy and (Anti) Aging: Mixed Blessings. 1 ed. Springer Netherlands; 2013. p. 105-18.

(35) Schoonheim case. Nederlandse Jurisprudentie 1984 Nov 27;(1985, 106).



# Perhaps the subject of the questionnaire was too sensitive: Do we expect too much too soon?

## Wishes for the end of life in Huntington's Disease – the perspective of European physicians

Suzanne J Booij
Aad Tibben
Dick P Engberts
Raymund AC Roos

Opinion piece.
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In The Netherlands euthanasia and physician-assisted suicide (PAS) have been legal since 2002 albeit under strict conditions.[1-3] This enabled patients who were suffering from a neurodegenerative disease, such as Huntington's Disease (HD), to make plans for the future. In our out-patient clinic specialized in HD we have experienced an increase in the number of requests for euthanasia and the use of advance directives. Further investigation has revealed that many HD patients and identified gene carriers do have some ideas about what they want regarding the end of their life.[4] We then wondered if physicians in other European countries, who are familiar with HD, have made similar observations and whether they are willing to discuss the subject with their patients. We were also interested in the actual content of the patients' wishes and what the physician's reaction is to these or related questions.

In an attempt to gain insights into the European perspective, we developed a questionnaire which we sent to participants/investigators/physicians who participate in the European Huntington's Disease Network (EHDN). 540 questionnaires were sent out in February 2013 and a reminder in April 2013 to participants in 17 European countries, all by email. The questions addressed the end of life options and aimed 1. to investigate if physicians know about these options in their country, 2. to investigate if physicians ever have conversations with their patients about their wishes regarding the end of life and 3. what their opinion is about the possibilities in their own country. In addition, some biographic and demographic data were collected.

Only 53 questionnaires were returned, i.e. a response rate of 10%. Another 10 respondents indicated that they did not want to complete this questionnaire or participate in this study, for different reasons: lack of time, lack of interest, not enough contact with HD patients or not willing to participate because of the nature of the topic. Due to the low response rate, we cannot draw any firm conclusions, but we will make some general comments and wish to speculate on the reason for the low response.

The responses were derived from physicians in 15 European countries. Forty respondents were neurologists; just over half considered themselves to be religious. Most respondents answered that advance directives concerning: not to resuscitate (DNR), treatment limitations or the appointment of a representative are valid in their country.

Half of the respondents, representing 12 European countries, reported that their patients do express wishes for the end of life. These involved all aspects of care, administration of fluid and food, admission to a hospital or nursing home, DNR and euthanasia and PAS in countries where it is legal (The Netherlands, Belgium, Luxemburg and assisted suicide in Switzerland). In the majority of cases the patient or a relative took the initiative to discuss the wishes with the physician. In a minority of cases the physician actively asked the patient about his/her thoughts and feelings about the end of life. Physicians reported that the reasons for having these wishes were related to fear of future suffering, having witnessed suffering in a relative, and the fear of loss of control, independence and dignity. Most patients were in the early or advanced stages of the disease when the discussion about their wishes for the end of life took place.

Considering the management options for the end of life some respondents reported that "quality of life could also mean quality of dying, and thus, euthanasia or PAS": several other respondents indicated that their focus is on quality of life. "I am against any kind of assisted suicide as this is a reflection of giving-up, or an example of a missed diagnosis. It is far from my professional attitude to let an event like that pass" and "I'll fight for the health of my patients and their quality of life. I'll never take someone's life." For these respondents, assisting in euthanasia or PAS is not an option.

Several respondents indicated that they would like to see management options for patients being expanded in the future, because patients do have end-of-life wishes. It would seem, however, that others ignore this fact. "It should be recognised by our law" and "I hope that our law could change. They ignore the problem, but these patients exist" and "there is a strong opposition from some sectors of the society, such as the church." One physician expected "there would be a decrease in the number of suicides in the HD population if wishes for end of life could be expressed and euthanasia or PAS legalised."

Reviewing the responses we did receive, we can only speculate about the reasons for not being willing to participate in this study. From the questionnaires returned, we can deduce that many physicians probably do get questions from patients about their choices for the end of life, treatment limitations and retaining a certain quality of life. Perhaps the nature of the questionnaire was too delicate, although we did not direct our questionnaire towards euthanasia and PAS solely, but towards wishes for the end of life in a broader perspective. It may be that talking about euthanasia, PAS or other life-limiting actions is still difficult in parts of Europe, but we feel that our questionnaire was drafted in such a way as to invite physicians to talk about wishes for the end of life in general.

A second reason could be that physicians perceived it to be not important or irrelevant to their practice to answer. Other studies asking about practices surrounding the end of life in general reported higher response rates.[5;6] None of these studies were directed towards a specific disease. A reason for a higher response rate in one study could be that physicians in this study were selected because of their involvement in the treatment and death of a specific patient instead of our choice to send the questionnaire to every physician treating patients with HD and asking general questions about the end of life. Some physicians may consider that it is not part of their professional duty to talk about care at the end of life. But, is it not the responsibility of the physician to care for his patient until the end of the disease? Is caregiving not always synonymous with quality of life which can also mean quality of dying for an individual patient?[7-9] As some physicians say: “I am always glad when the patients are admitted to a nursing home, then I do not have to discuss the end of life issues.”

A third reason might be that physicians receive many requests by email. However our study group was not randomly selected, but approached because of their involvement with HD, their participation in HD research and their membership of EHDN. From this point of view it is even more remarkable that response rate was so low, which could support our hypothesis that the issue might have been too delicate.

Ways to improve response rate could be for example to hand out the questionnaire during the international congresses on HD and ask physicians to fill in the questionnaire during the congress.

From our studies we concluded that HD patients do have wishes and do want to talk about their wishes with their physician.[4] Talking about thoughts and wishes for the end of life and talking about quality of life and all that quality of life encompasses can be very helpful and reassuring for patients.[10] As patient’s autonomy attracts an increasing amount of attention in the public debate, choosing not to be treated will become part of doctor-patient conversations.[11] Studies have shown that families rely on physicians to help in the decision-making process.[11] A research paper reported that patient autonomy could even be transferred to the physician when the patient was no longer competent according to the family.[12] Furthermore, other research has indicated that some form of acceptance and regulation of euthanasia and physician-assisted suicide is gaining increasing support from the general population in most western countries.[13]

Are we expecting too much at this moment by drawing attention to this topic or should doctors prepare themselves better for this conversation with their patients? Thus are we expecting that we can ask these kind of questions to physicians in other countries, because we are so used to talking about this topic and discussing these wishes in The Netherlands? Is the time not there yet, is it too soon to ask questions about this topic to physicians in other countries, should we wait a couple of years? Or should doctors consider taking care of their patient in all aspects of their disease, the patient's welfare and best interests, including quality of dying, as part of their job?

In our opinion doctors will receive questions from patients about this topic more frequently in the near future. Other studies already concluded that physicians with training in palliative care were more inclined to make end-of-life decisions.[6] We do not suggest that other countries should legalize euthanasia or PAS in the near future. Nevertheless, the first step towards helping patients is to make the topic of end-of-life wishes open for discussion. We think that the awareness that HD patients have end-of-life wishes and the awareness that a physician and patient (together) should make end-of-life decisions, can increase the realization that this is indeed a very relevant topic for every physician treating patients with HD and thus to respond to these kinds of questions.

## Acknowledgements

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## REFERENCES

(1) http://www.euthanasiecommissie.nl. 2014.

(2) Onwuteaka-Philipsen BD, Brinkman-Stoppelenburg A, Penning C, de Jong-Krul GJ, van Delden JJ, van der Heide A. Trends in end-of-life practices before and after the enactment of the euthanasia law in the Netherlands from 1990 to 2010: a repeated cross-sectional survey. Lancet 2012 Sep 8;380(9845):908-15.

(3) van der Heide A, Onwuteaka-Philipsen BD, Rurup ML, Buiting HM, van Delden JJ, Hanssen-de Wolf JE, et al. End-of-life practices in the Netherlands under the Euthanasia Act. N Engl J Med 2007 May 10;356(19):1957-65.

(4) Booij SJ, R+¦dig V, Engberts DP, Tibben A, Roos RAC. Euthanasia and Advance Directives in Huntington's Disease: Qualitative Analysis of Interviews with Patients. Journal of Huntington's Disease 2013 Jan 1;2(3):323-30.

(5) Cohen J, Bilsen J, Fischer S, Lofmark R, Norup M, van der Heide A, et al. End-of-life decision-making in Belgium, Denmark, Sweden and Switzerland: does place of death make a difference? J Epidemiol Community Health 2007 Dec;61(12):1062-8.

(6) Lofmark R, Nilstun T, Cartwright C, Fischer S, van der Heide A, Mortier F, et al. Physicians' experiences with end-of-life decision-making: survey in 6 European countries and Australia. BMC Med 2008;6:4.

(7) Daher M. Ethical issues in the geriatric patient with advanced cancer 'living to the end'. Ann Oncol 2013 Oct;24 Suppl 7:vii55-vii58.

(8) Gillett G, Chamberlain J. The clinician's dilemma: two dimensions of ethical care. Int J Law Psychiatry 2013 Sep;36(5-6):454-60.

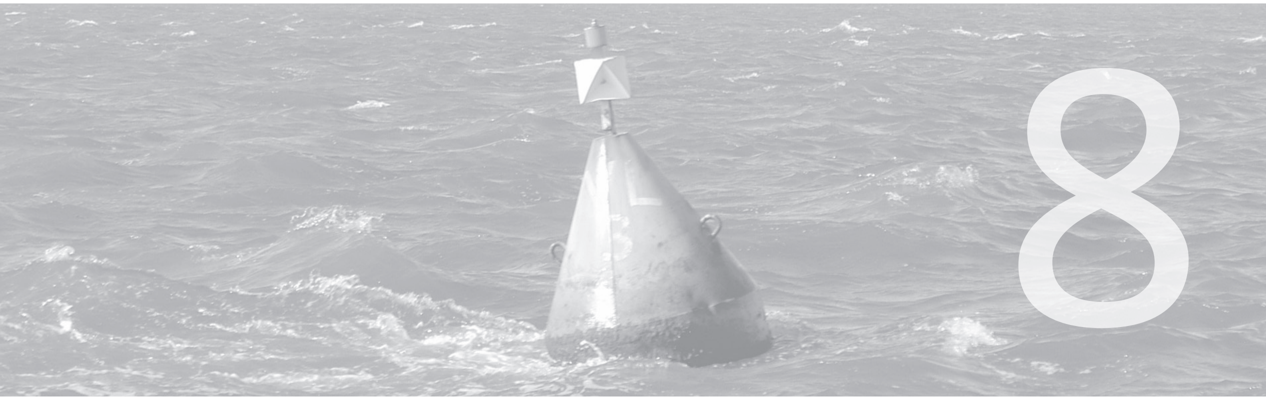
(9) Borgsteede SD, Deliens L, Graafland-Riedstra C, Francke AL, van der Wal G, Willems DL. Communication about euthanasia in general practice: opinions and experiences of patients and their general practitioners. Patient Educ Couns 2007 May;66(2):156-61.

(10) Booij SJ, Engberts DP, Rodig V, Tibben A, Roos RA. A plea for end-of-life discussions with patients suffering from Huntington's disease: the role of the physician. J Med Ethics 2013 Oct;39(10):621-4.

(11) Billings JA, Krakauer EL. On Patient Autonomy and Physician Responsibility in End-of-Life Care. Arch Intern Med 2011 May 9;171(9):849-53.

(12) Monteiro F. Family reliance on physicians' decisions in life-sustaining treatments in acute-on-chronic respiratory diseases in a respiratory intensive care unit: A single center study. Respir Care 2013 Aug 20.

(13) van der Heide A. Assisted suicide and euthanasia. Handb Clin Neurol 2013;118:181-9.



# 8

# General discussion

## Focus of this thesis

Even before the media attention for this topic, euthanasia and end-of-life care in Huntington's Disease (HD) seemed to attract the attention of patients and families and became a subject several patients wanted to discuss.

Therefore the aims of this thesis were I) are end of life wishes present in patients with Huntington's disease and known gene carriers, II) are patients willing to discuss these wishes, III) what is the content of these end-of-life wishes; IV) do physicians actually discuss wishes for the end of life with their patients and V) how do physicians cope with these issues and questions. In this final chapter an overview of the main research findings is presented and discussed.

Subsequently a few key issues on the law on euthanasia and physician assisted suicide (PAS) in The Netherlands are reflected upon; advance care planning and the usefulness of advance directives with special attention for a progressive neurodegenerative disease such as HD. We focus on two key signs of HD, namely cognitive decline and psychiatric symptoms, which present specific difficulties when thinking about euthanasia or PAS and when trying to fulfil a patient's wish.

## Main findings

Following a study of the recent literature, the law on euthanasia, the development of the law and the applicability of euthanasia in HD is reviewed (**chapter 2**). The development of criminal court decisions in the context of social history, and the increasingly recognized patient rights and right to decide about one's own life without leaving it up to the physician, led to the implementation of the 'Law on review on the ending of life on request and assisted suicide' in 2002. The fact that HD is an inherited disease and persons at risk can be tested early in life with a accuracy of nearly 100% led to the situation that patients want a say in their future, their life and therefore also their death. The exploration of the legal possibility of euthanasia in HD patients showed that euthanasia is very well possible in patients who are affected by this neurodegenerative disease despite the fact that all patients will suffer from dementia at some point in time. An advance directive, advance care planning and repeated conversations with the physician are of great importance. The importance of keeping the shared opinions and values -and their notification in the patient's medical record- up to date seems somewhat neglected by physicians sometimes.

## The wishes of patients

The results of one of the main questions of this thesis and the reason for starting this study are described in **chapter 3 and 4**. During the interviews with an unselected group of patients we discovered that asking questions about the end of life is not considered a burden. In general patients reacted positively and were willing to comply. One of the main findings is that there are probably more patients who think about the end of their life than is currently known to physicians. Although the qualitative approach only let us discover themes, a remarkable finding was that patients think about the end of life, but have not always talked to their physician about it. Reasons for this restraint were that end-of-life wishes were not yet clear to the patient and that the patient only had some general ideas about what he/she wanted to prevent of avoid, but these thoughts were not explicit yet. Furthermore and possibly because of this somewhat ambivalent attitude towards the subject, there is a lack of knowledge about the legal and practical possibilities and requirements.

The themes described in **chapter 3** about the presence of wishes and the lack of knowledge were further explored with a qualitative study. Possible influences of patient characteristics and disease characteristics which were not detected by our qualitative study were further investigated. These characteristics include age, gender, religion, education and disease stage and were investigated by the use of a questionnaire. The results are described in **chapter 4**. Out of 249 patients that were approached and received a questionnaire, we got only one explicit negative reaction. One-hundred-and-thirty-four patients returned their questionnaire and we did not receive a response from 114 patients. Our research showed that thinking about the end of life as HD patient or as a premanifest gene carrier is not restricted to advance disease state, but is something of all stages of the disease. Patients have thoughts about their end of life. These thoughts are sometimes just confined to wishes about how to organize care in the future, but many times thoughts are directed towards how to have a say in the end of life or to determine one’s own end of life. Presence of wishes was not correlated to any demographic characteristic. The only patient characteristic that was correlated with wishes for the end of life was knowledge about the disease and future prospects based on experiences with family members who had or have this disease.

There was only a minor group of patients who have an advance directive. We can only speculate about the reason why, but derived from our qualitative research, a hypothesis could be that thoughts about the end of life are maybe just thoughts at some point in time and not yet conclusive enough to be written down and documented.

## The role of a physician

The results and interpretations of the qualitative interviews with the physicians are presented in **chapter 5 and 6**. The results showed that the physicians are open and willing to talk about the subject, but leave it up to the patient to enter into such a conversation. The requirements of the law are usually known but remain somewhat theoretical. The use of advance directives, the task a physician has when exploring wishes for the end of life with patients with a neurodegenerative disease and the implementation into everyday practice proved to be difficult. During analysis of the interviews some themes were recognized that can only be discovered with qualitative techniques, such as the evaluation of suffering of a patient, how to handle response shift and the different opinion upon a professional's task when entering into a conversation about the end of life.

These findings gave the opportunity to suggest the use of an instrument to help a physician in guiding the patients. The instrument is the use of a letter of intent. In that letter a physician writes down the request of the patient and declares it to be his intent to help the patient in due course with the fulfillment of the wishes. The intent shows that a physician will take the request serious, without promising to reach the result wished for. An important part of the letter consists of the description of the wishes of the patient and the frequency upon which the conversation will be repeated. The document can also be used to educate the patient further upon the requirements of the law; to make sure the patient does not assume things are taken care of after writing and filing an advance directive. The letter of intent also reassures an active role of the physician. Because even though applicable situations are recognized, especially by nursing home physicians, the advance directive is sometimes declared inapplicable. In these cases the physician is not able to tell if the situation the patient is in now, is the one mentioned and referred to in the advance directive. Secondly the physician is not convinced of the suffering and of the fact that this is really what the patient wants. Taking a more active role and entering into conversations about the end of life can make complying with an advance directive easier. It can prevent suffering by the patient.

## European physicians familiar with HD asked about patients' wishes for the end of life

The last part of the study was to explore if physicians in other European countries also perceive that end-of-life wishes are present in patients with HD and if physicians discuss these wishes with HD patients (**chapter 7**). We developed a questionnaire and sent it to European physicians, members of the European Huntington Disease Network (EHDN). Unfortunately the response rate was very low. We can only speculate on the reasons for this low response rate. One can be that physicians are tired of questionnaires and decided not to answer. A second can be that this is not a topic that is of importance for HD patients in their country. Thirdly, it is possible that physicians try to avoid this topic, because of its illegal status in their country or because they think it is too difficult a topic to enter into with patients.

From the returned questionnaires we can draw only minor conclusions. We received an answer from every European country participating in EHDN. This gives us a fairly general insight into the possibilities for the end of life and possibilities to talk about, write down and respect end-of-life wishes for HD patients in each country. The responses showed us that in other European countries wishes for the end of life exist in patients and patients discuss these wishes with their physicians. We concluded that as physicians we cannot ignore this topic and with more and more patients practicing and demanding their autonomy we will probably be increasingly confronted with discussions of this kind.

## Euthanasia in advanced Huntington's Disease

Reports on performed euthanasia and PAS increase each year in The Netherlands.[1] Two main categories of diseases attract the attention of the general public and politics in the debate about euthanasia in recent years: psychiatric diseases and dementia. The Regional Review Committees for euthanasia (RRC) publish their findings and core cases every year and pay extra attention to euthanasia in case of an underlying psychiatric disease or dementia more frequently.[1]

Huntington's Disease (HD) has features of both, psychiatric signs and symptoms and cognitive decline leading to dementia. In 2012 an interesting case was reported to the RRC, that attracted a lot of attention of the general public and in the media. In this case a general practitioner reported to the RRC about euthanasia in a patient diagnosed with HD. Six years earlier the patient had talked to her general practitioner about euthanasia for the first time. She signed an advance directive. She also talked to her family about her wishes and appointed her husband as her legal representative in case she was not able to express her wishes anymore. In the years that followed the patient and her physician did not regularly talk about euthanasia or the advance directive. Four years after the first conversation the general practitioner wanted to talk with her about euthanasia again but the patient ended the conversation quickly saying she didn't want an injection at that time. Two years later communication was hardly possible anymore. During her last visit to the outpatient clinic of her neurologist the patient uttered only one word, 'euthanasia'. When the subject of euthanasia was brought up by the general practitioner later the patient did not react or respond. Finally her husband asked for euthanasia and the request was honored. The RRC concluded that since several years had passed between the first conversation about euthanasia and signing of the advance directive and the impossibility of conversation six years later, the general practitioner should not have come to the conclusion that the request for euthanasia was made voluntary and well considered. Secondly, although the patient had declared, also in writing, that admittance to a nursing home would be unbearable for her, it seemed the patient was reasonably content during the days she spent in a nursing home. Thus the physician could not conclude that the patient was suffering unbearably.[1]

In that same year a second case of euthanasia in a patient with advanced dementia and based on an advance directive was reported to a RRC. In this case the RRC concluded that the euthanasia was carried out within the limits set by the law because the patient spoke repeatedly about euthanasia with her physician, could well describe what unbearable suffering meant to her and made it clear to her physicians and others, both verbally and non-verbally, that she wanted to die.[1]

The difference between these two cases was the possibility of conversation between patient and physician and the fact that conversation about possible euthanasia occurred only a few times in the case of the first patient and repeatedly in the second. The comments of the RRC in these cases provide further clues and help for physicians discussing euthanasia with patients suffering from dementia, and it helps deciding if possibilities exist for euthanasia in case of dementia, based on an advance directive.

## What euthanasia actually beholds: anticipating the future

As described briefly in **chapter 2** the debate about euthanasia in The Netherlands has been going on since the 1960s. In the early eighties some physicians started to report that they had performed euthanasia and the chief public prosecutor decided to bring some of them to court to produce clarity and judicial rulings with which to go forward. Several key cases have been brought to the Hoge Raad (Supreme Court) since then, clarifying the requirements of due care and setting boundaries as to the scope of the possibilities physicians have.[2]

The first ruling where a verdict is asked of the Supreme Court in case of death on request is the verdict in the case of dr. Postma in 1973.[3] This was the earliest case in the development of Dutch euthanasia law, but was one of ‘private’ assistance. The physician who carried out the euthanasia was not the patient’s treating physician but the daughter of the patient. In this case the father of the daughter/physician died after a long period of suffering and the mother/patient decided that she did not want to die that way. The mother/patient suffered from several somatic illnesses, was admitted to a nursing home and eventually also became depressed. The depression proved to be hardly treatable and rehabilitation failed. The mother/patient tried to commit suicide and in the end the physician/daughter decided to help her mother.

There are many differences between this case and the requirements of due care that are requested now-a-days, which are elaborated in rulings of the Supreme Court in the years that followed.

The second and most important case to set the framework of requirements of assisting a patient to die on his request is described in **chapter 2** and is the case of dr. Schoonheim in 1984. In this case the Supreme Court ruled that euthanasia could be legally justifiable under specific conditions.[4] We described these conditions in chapter 2.

A third interesting case when reviewing the possibilities and the boundaries of the possibilities provided for in the law, was the case of senator Brongersma.[5] During the last years of his life, Brongersma was diagnosed with several illnesses and discomforts, all by themselves not life-threatening, but negatively influencing his quality of life. He had a depressed mood, but was not diagnosed with a depression. He repeatedly asked his general practitioner to help him to die and in the end the general practitioner did. The Supreme Court however ruled that the general practitioner did not act within the scope of the limits set by the court rulings of the past years. Reason for the rejection of the appeal to necessity was that the specific conditions that make euthanasia justifiable do not apply to situations where the suffering is not predominantly due to a medically classified illness, either somatic or psychiatric. One of the requirements is that the physician should act according to scientific medical views and insights and ethical standards of the profession. In case of a request for euthanasia without a medical classified illness a physician does not act according to these requirements, because the expertise of a physician is limited to the medical domain and does not extend to more general questions of life and death.[6]

Reflecting on these three important cases and the considerations of the Supreme Court and the development of the euthanasia law and practice in The Netherlands we can ask ourselves what the meaning of euthanasia actually is for patients. The word euthanasia comes from the Greek word 'εὐθανατος ', meaning a good death. The patients in the cases of dr. Postma and dr. Schoonheim both anticipate upon future suffering and asked for a good death, without further suffering. When thinking about the request for euthanasia or PAS, could we not come to the conclusion that euthanasia or PAS mostly is about anticipating and avoiding future or imminent suffering? Because when somatic suffering is present, a physician has its options, ways and tools to relieve that suffering. When a patient is in pain, is short of breath or uncomfortable physicians have medication to relieve suffering, without talking about euthanasia or PAS. Most physicians can use palliative sedation to relieve signs and symptoms of disease. Sometimes these kind of treatments may lead to an earlier death, but that possibility was not intended and the treatment was not started to hasten death.

Thus when somatic suffering can be relieved, why do we need euthanasia and PAS? In our opinion we need euthanasia and PAS because they represent an option for patients to avoid suffering, to not come to the stage where suffering must be relieved for example by inducing a reduced consciousness and to provide a possibility to stay in control of one's own life. Thus to avoid suffering and stay in control, a patient anticipates the future.

If we accept this conclusion that euthanasia and PAS are to anticipate future suffering, it brings us to several interesting topics. First the advance directive. The advance euthanasia directive is developed to anticipate future suffering. But are we then anticipating to the anticipation of future suffering? Secondly, the meaning of the second article, second paragraph of the euthanasia law in The Netherlands, stating that in case of an advance directive, the other requirements of due care are accordingly applicable. And a third topic can be the wish for euthanasia in case of dementia. Is a patient able to anticipate future dementia?

## Anticipating future suffering

Suffering is understood and interpreted differently in different contexts. In palliative care the prevention of suffering and the relief of suffering is a major goal.[7] The origin of suffering is different for patients suffering from different diseases. For cancer patients and patients with heart failure suffering is related to somatic complaints. For patients with amyotrophic lateral sclerosis suffering beholds fear of suffocation and dependency.[8] For HD patients a reason to ask for euthanasia is fear of dependency, dementia and future signs and symptoms of the disease, usually after having witnessed a family member (**chapter 3 and 4**). For laypeople the concept of suffering usually conjures images of patients with pain.[9] Suffering is seen and dealt with in many different ways and people who suffer are understood in different ways, for example: suffering is part of life, or people who suffer are without hope, waiting to die, or that allowing people to suffer is wrong and the ending of suffering is a right.[7] The way we perceive suffering as individuals will probably also decide how we cope with suffering and how we anticipate on future suffering. The avoidance of suffering has been used as an argument for the legalization of euthanasia and physician assisted suicide.

Competent patients are seen as autonomic individuals, capable of making their own medical decisions, including decisions about medical treatments and also death, instead of patients listening to physicians and following their physicians' decisions.[10;11] Dignity is associated with independence and preserving one's intellectual powers.[12;13] Recognizing each competent individual as capable of making their own autonomous decisions is one of the key constituents of modern Western medicine. A patient can make his own decisions, whether the doctor agrees with the decision or not. Autonomy and dignity as concepts led in the medical domain to the fact that patients wanted a say in their life and death, wanted to control the end of life and this process became part of self-identity. Deciding when it is time to die became part of dying in accord with the individual's personal way of living. The fear of losing dignity and autonomy are earlier described as reasons for asking for euthanasia in time.[12;13] Furthermore, another study showed that when asked, depending on others may be perceived by older persons as problematic and socially as being a 'burden'.[12]

This fear can only be understood as fear for the future, fear for future dependence and loss of dignity, fear of not being able and capable to make decisions anymore. But if this fear for these future possibilities is a reason to ask for euthanasia, we can come to the conclusion that asking for euthanasia means anticipating future suffering.

## Advance directives and euthanasia in the Dutch law

Euthanasia based on an advance directive remains one of the most debated topics of the law since the law passed in 2002. The law on euthanasia and PAS and the legalization of those acts by a physician states in the second article paragraph 2 that euthanasia and PAS are possible, based on an advance directive, replacing a direct request, taking the other requirements of due care, which are accordingly applicable, into account. But what does accordingly applicable means when we describe euthanasia as anticipation to future suffering and an advance directive as anticipating to the anticipation of future suffering?

As described extensively in literature the first requirement for a physician to explore is if the request is voluntary and well considered.[14] When a physician discusses the advance directive with the patient at the moment it is filled out, he can explore these requirements.[15] This may not always be done. As we described in **chapter 3** patients are not always aware that when following the law, it is the physician who is leading in the process that leads to euthanasia. It is the physician who has to be convinced that there is no other possibility to relieve suffering and no other solution in this case than euthanasia. Patients are not always aware that the physician must be part of the process of determining wishes for the end of life and filling out an advance directive.

When the advance directive is not discussed when it is filled out, the physician can have the opportunity to discuss the advance directive when it comes to his attention that an advance directive is present. The physician thus explores and knows the will of the patient, knows that the request is well considered and voluntary at that moment. But how can a physician decide if euthanasia or PAS still is the wish of a patient, for example 10 years later? Parliament decided that the advance directive reflects the will of a patient, the past will of the patient is ‘translated’ into the present. In order to know and understand the will of a patient, the advance directive must be explicit and signed and dated by the patient.[16] Parliament also stated that if all requirements of due care were met, a physician can still decide not to go forward with the requested act, because of well-founded reasons, for example the state of medicine now offers more possibilities that those that were present at the time of signing the advance directive. The physician is expected to imagine what the will of the patient would have been in these changed circumstances.[16] This example is a somewhat strange one and shows overlap with the fourth requirement, but was meant to support the reasoning of Parliament that a physician can always decide not to go forward with the requested euthanasia. The fourth requirement of due care states that no reasonable alternative exists in light of the patient's situation. If new treatment possibilities exist , reasonable alternatives could also be present and thus the other requirements are not accordingly applicable in such a situation. We conclude that this example represents some of the difficulties that arise with this paragraph of the law.

## 'Accordingly applicable' or applicable in a corresponding way

If we assume a physician is able to know the will of the patient (now and in the future), we turn our attention towards the other requirements, which are 'accordingly applicable' or applicable in a corresponding way. The second requirement is the requirement of unbearable suffering without prospect of improvement. Severely demented patients are generally believed not to suffer from their condition, according to the Ministers of Justice and Health and according to current scientific standards.[2;12;17] This second paragraph was even considered an empty shell, especially for patients suffering from dementia, for whom it was largely intended, because of the fact that the other requirements could not be met.[2] Nevertheless parliament stated that the advance directive can serve as a guidance for a physician when confronted with a patient who is no longer able to express his will. A physician could and should however always make his own judgment.[16]

Knowing the position of Parliament we turn our attention towards the phrase 'accordingly applicable' or applicable in a corresponding way. Parliament does not take a position upon this phrase other than that a physician should consider the possibilities and make his own judgment. But when should the other requirements be accordingly applicable? If we accept that euthanasia is anticipating future suffering, the suffering must be described in the advance directive. The patient must have anticipated what he feared and expected that to be unbearable. Anticipating the future does not take the possibility of response shift into account. We wrote about response shift in **chapter 6**.

Furthermore what does accordingly applicable mean in the context of the requirement that 'the physician informed the patient about his situation and his prospects' when communication is no longer possible? The patient can be informed when he writes down his wishes and when he draughts his advance directive. If the situation changes in the following years, the advance directive can be adapted to this new situation. But what if the advance directive can no longer be adapted, because the patient is no longer aware of his situation or no longer capable to express his awareness of his situation, as is possible in advanced HD?

Parliament again did not take a position upon the difficulty with this problem, but I believe that some solution is provided in the explanation of the use of an advance directive. The past will of the patient is 'translated' to the present. By talking with the patient regularly about his situation and prospects, a physician is able to know the will of the patient and the consistency of the will. And if the situation changes he can try to extrapolate the will of the past into the future and decide what this new information would change about the situation and the prospects.

The same explanation holds for the requirement that patient and physician together should come to the conviction that there is no reasonable alternative in light of the situation. The physician and patient have talked about the situation repeatedly and together decided that, in due course, euthanasia or PAS was going to be the only possible solution. When communication is no longer possible, it is difficult, not to say impossible, to come to this conviction together that euthanasia or PAS is the only possible solution, but again the physician can follow up on his experiences from the previous period. In this situation the information of relatives can also be informative and important, even though a physician cannot act solely on the basis of information from relatives.

The fifth requirement of the law states that an independent second physician must visit the patient and give a written opinion on the due care criteria. It seems difficult to evaluate these criteria for an independent physician, when he meets the patient at a moment in time when communication is hardly possible or dementia has progressed to a stage where a conversation about the voluntary and well-considered wish and the content of unbearable suffering is hardly possible because of cognitive deficits.

A possible solution lies in the relationship with the patient and the anticipation upon the request by the first or 'performing' physician. This second physician maybe less able to translate the past will into the present when he meets the patient at a late stage in the process. However when physician and patient anticipate a future request for euthanasia they can anticipate on this requirement as well by asking the independent physician to visit the patient early and give his written opinion in an early stage. At present, current rulings of the RRC state that the report of the second physician has limited shelf life and an early consultation cannot replace a later one. However if this second physician repeatedly visits the patient a firm and thorough dossier will be available. With this dossier a RRC can follow the line of reasoning of both the first and second physician. A RRC can evaluate and make a substantive assessment of the performed euthanasia or PAS and the criteria of due care.

If we read the requirements very strictly, one may argue that the accordingly applicable requirements only apply to the suffering without future prospect of improvement and the voluntary and well-considered request. The requirements of informing the patient about his situation and prospects and being convinced there is no reasonable alternative in light of a rapidly changing medical science and rapidly increasing treatment possibilities seems difficult to adhere to and to anticipate in the future. Secondly the consultation of an independent second physician who should give a written opinion on meeting the criteria also seems difficult to make ‘accordingly applicable’ when communication is no longer possible. On the other hand, looking at the explanation of Parliament and reasoning in light of their thoughts, Parliament did provide space for a physician to interpret and judge every case individually. Anticipating and extrapolating the past wishes of a patient into the present situation, anticipating future suffering and a future wish for euthanasia by preparing everything in advance, when trying to find an acceptable solution for a patient, a physician enters a domain where he can act, but where the law is stretched to the limit. Solutions are available, but must be carefully weighed and extensively discussed with all parties involved. Reaching the limits of the law, stretching these limits and being aware of this process can only be allowed when the process and the action are checked, balanced, controlled and judged upon. We find ourselves in a transition from law towards ethics.

## Recent developments in the debate about euthanasia in case of dementia, based on an advance directive

The Health Council of The Netherlands concluded already in 2002 that dementia alone is not enough ground for euthanasia. Main difficulties that were recognized and described were 1. the difficulty to decide when a patient is incompetent and lost its decision making capacities to such an extent that the present acts of will should be ignored in favor of the advance directive and 2. if patients with dementia actually suffer unbearably and hopelessly. A third recognized but not mentioned dilemma by the Health Council is the absence of a physician-patient relationship and conversation about the wishes when a patient is in an advanced stage of dementia. Several groups then proposed to shift attention towards the early stages of dementia. However, as prof Hertogh stated, a patient suffering from dementia has two options: “either deny what is happening and loose contact with reality or face the reality and realize that he/she is losing oneself.”[18;19]

In 2012 the Royal Dutch Medical Association (KNMG) published its sharpened position in the matter of euthanasia in dementia, based on an advance directive.[20] The KNMG stated that without communication euthanasia is impossible. The physician of the patient and the independent second physician should both evaluate the request for euthanasia and communicate with the patient about his wish, suffering and alternatives. The sharpened position of the KNMG ‘adds’ to the law the requirement of communication whereas the law only states that an advance directive can replace the expressed request.[21] As discussed earlier we recognize the difficulties with advance directives and the phrase ‘accordingly applicable’ in the law, however we feel that with this position the KNMG ignores the possibilities provided for in the law, for example role and function of the independent second consultation of a physician, which is to investigate and form his opinion on requirements of due care and not to agree with the performing physician or consent. We also proposed a possible solution in the previous paragraph.

But euthanasia in case of dementia based on an advance directive does not only concerns communication, the voluntary and well considered request and a discussion about suffering. It is also about the future. When a patient draws an advance directive and states that he wants euthanasia in case of dementia, as is the case for many persons in The Netherlands, the person anticipates the future. The person anticipates future suffering, caused by dementia. However it seems almost impossible to anticipate future dementia and future suffering from dementia. Therefore it seems impossible to draft an advance directive stating a person wants euthanasia in case of dementia. It seems impossible to anticipate what dementia beholds for an individual, what signs and symptoms a person will have and if a person actually suffers from the dementia. On the other hand, our investigation of the accordingly applicable requirements of due care and the proposition on how to cope with these requirements, provide a solution for physicians to cope with advance directives for euthanasia in case of dementia. To anticipate means involving a physician early in the process, as we already stated and stressed earlier.

## Huntington’s Disease: the centre of a triangle of somatic, psychiatric and cognitive disease

Returning to the main subject of this thesis we incorporate our views upon the anticipation of future suffering and the applicability of advance directives into our findings about patients with Huntington’s Disease (HD). As mentioned by the Health Council and several authors HD is different compared to most other diseases, both somatic and neurodegenerative because patients can actually anticipate the future much better. Patients know what their future will be like for two reasons, their genetic status and their experience with a family member, even though of course every individual has its own course. Patients know for sure that they will have problems with cognition leading to dementia, and that they will someday be dependent on others for every day care, for most of them leading to admittance to institutionalized care. The possible suffering includes many different factors, both somatic, cognitive and psychiatric. For HD patients suffering is not only anticipated as a possibility, most patients know for certain that some form of suffering will happen.

HD can be placed in the middle of a triangle of the somatic, the psychiatric and the cognitive aspects of disease or at the crossroad of somatic disease, psychiatric disease and dementia. On the other hand, literature states that HD is different, because of its inherited genetic cause and the extensive knowledge about disease course, but is that really the case? How is HD different from many other diseases?

With HD, thinking about the future, having a say in one’s life and death, deciding when quality of life has reached rock-bottom levels, it is all anticipating future suffering as with many other diseases. When a patient suffers from a somatic disease such as cancer or ALS or any other disease, there is some form of suffering at that moment and the patient looks into the future to anticipate what suffering can be expected or feared, that makes the suffering unbearable. HD patients do not look at their situation from a healthy perspective, they are already ‘patients’ in a certain way after a genetic test. Even though they may not have symptoms yet, they know they will develop this diseases in due course. Their perspective is somewhat similar to the perspective of patients with somatic diseases. Patients diagnosed with a somatic disease are patients from that moment on. They experience life and live on with their disease and their symptoms. The perspective of these patients is one of living with a certain disease and sometimes adapting to a new way of living. HD patients know, after a genetic test, that they have to adapt to a different life as well.

Furthermore, after the diagnosis of being a gene carrier for HD, these persons already suffer to some extent, as can be derived from the fact that the rate of suicide is increased just after a confirmative test result and just after a confirmation that signs and symptoms of the disease are present.[22-26]

This perspective of HD patients denotes a key difference between persons making an advance directive in case of dementia and HD patients, looking at their future, with knowledgeable perspectives. HD is usually mentioned as an example disease for other neurodegenerative diseases (dementias), but because of its many similarities with somatic diseases, it should both be looked at as a somatic disease and as a (form of) dementia.

A similarity between dementia and HD is the factor time. Although a diagnosis of dementia is more difficult, based on a complex spectrum of clinical criteria, neuropsychological and radiologic results, after diagnosis usually several years lie ahead. When a patient faces reality, there is an opportunity and there is time to prepare an advance directive and possible euthanasia request. Response shift will always be part of the process of disease, but only time can tell if a patient is able to cope with the disease and all its aspects and if response shift occurs or if a patient decides that he can cope to a certain point and after that point makes a choice to end life. When there is time at the hands of both physician and patient, progressive cognitive decline can be followed and anticipated on.

On the other hand, with time on the hands of a patient and his physician, when talking about wishes for the end of life with patients suffering from a neurodegenerative disease, leading to dementia, the question can arise to what extent patients no longer capable of autonomous decision making can have a say in their future, medical treatment and death. This questions touches the ethical debate of autonomy, precedent autonomy, former preferences and the value of autonomy and of preferences.[12;27;28] These dilemmas have extensively been discussed in literature and cannot be discussed or solved in the scope of this general discussion. Instead we choose to stay close to the possibilities provided for physicians by Parliament and the solutions suggested in this thesis. Parliament decided that former preferences should be translated into present ones and in this thesis we stated that communication is the key to solving the problem of dealing with cognitive decline, psychiatric symptoms and signs and how to cope with response shift.

In summary a lot can be said about autonomy and the value of autonomy. For example when a person decides to end his life, in literature it is postulated that because autonomy is valuable, it needs to be respected. This is explained twofold. One explanation is that autonomy is part of a person and when the person is no longer there, neither is his autonomy. On this grounds, euthanasia and PAS should be rejected.[29]

On the other hand Parliament choose the solution that former preferences should be transferred and translated into present ones and people should be able to make their own decisions. This is based on the findings that, especially in early dementia, patients are able to tell about their experiences with the disease and make decisions accordingly.[17;18;30] When dementia progresses it becomes increasingly difficult to decide if the moment has arrived the patient mentioned in his advance directive; a struggle evolves between acting according to the patient’s individual dignity and having respect for his (precedent) autonomy versus acting in the patient’s best and current interests and taking care of the patient, when mental capacities have diminished to such an extent that a patient is no longer able to express his wishes.[12;27] Again time can be of help, to monitor cognitive decline and anticipate, for a patient, for a physician and for family members. The time can be used to talk about the experiences with the disease and the preferences of the patient on how to cope with the disease. Knowing these preferences and repeatedly talking about them provides the opportunity to respect the patient’s autonomy and translate his previous autonomous decision about his future into a present choice for medical treatment or death.

A second similarity is the probability of the absence of treatment and thus reasonable alternatives for euthanasia, at this moment and in the near future. Advance directives can be prepared and the other requirements can be accordingly applicable, including the extrapolating of past wishes into the present. When taking the wishes into account that are written down in an advance directive, the physician has knowledge about what suffering beholds for this individual patient. The physician must have come to the conclusion earlier and together with the patient and must be convinced that for the situation the patient is in now, no reasonable alternative exists.

The third arm of the triangle is that of psychiatry, part of HD. Psychiatric signs and symptoms represent their own difficulties, with regard to a request for euthanasia or PAS. These aspects are discussed hereafter.

## Psychiatry and euthanasia

Another key feature of HD is psychiatric symptoms and signs. The psychiatric symptoms and signs include a broad spectrum of characteristics, including depression, suicidality, irritability, apathy and anxiety. Less frequent symptoms include obsessive-compulsive behaviour, with rates reported up to 50%.[31]

The earliest case of euthanasia in a patient overwhelmed by mental suffering without an official somatic or psychiatric diagnosis was the case of dr. Chabot in 1994, which judicial ruling is still very important and up-to-date nowadays, even after the introduction of the 'Law on review on the ending of life on request and assisted suicide' in 2002.[32] In this case the Supreme Court stated that 'suffering must be abstracted from the origin of suffering to such an extent that the origin does not affect the extent to which the suffering is experienced. This does not alter the fact that, when suffering does not arise from a somatic disease, it is more difficult to determine the unbearableness of the suffering to which no prospect of improvement exists."

With this ruling the Supreme Court opened a door for euthanasia and PAS in case of psychiatric disease and suffering from a psychiatric disease. The Supreme Court concluded that to call for a statutory defence suffering is independent from somatic disease or being in the last stages of disease. Suffering is always a subjective experience and irrespective of the origin has a mental dimension.[32] The extent to which suffering from a psychiatric disease could be regarded as a reason for euthanasia or PAS presented problems for all parties involved during the development and draft of the law and afterwards, including Parliament. The minister of Health Care stated that euthanasia and PAS are limited to the context of medical treatment, which does not include psychiatric diseases.[33] On the other hand, the government stated elsewhere that when a patient with a psychiatric disease fulfils all the requirements of due care, the law could apply to these patients as well.[33] Looking at the way the law was constructed and formulated, the reasoning of the government was correct.

The RRC's pay extra attention to this subject in their annual reports.[1] Several reasons for heightened precaution in case of euthanasia in psychiatry have been described. First there is the possible influence of the disease itself on the request. Is the request a voluntary and well-considered request in itself or is the request the result of symptoms of the disease, for example suicidality in case of a depression. Secondly, suffering from a mental illness can be more difficult for others to understand and to recognize (**chapter 6**).

And thirdly, the controversy and sensitivity of these problems in society or with the public in general.[33 (page 18)]

### Case of a wish for euthanasia in HD in the context of psychiatric symptoms

To illustrate some these problems we describe a case of a HD patient with a wish for euthanasia.

#### Case 1

The patient is a male between 60-70 years old. He has had signs and symptoms of HD for several years. He talked about euthanasia with his general practitioner, neurologist and psychologist repeatedly in the last couple of years and made an advance directive. An independent second physician already visited him several times. He could well describe his experiences with the disease and could describe what suffering meant for him. Keeping his ability to leave the house whenever he wished, being independent and having a sense of freedom were very important factors for him. These factors all came together in his car and the possibility to drive a car. He was aware of the fact that HD in his case meant psychiatric symptoms such as irritability, aggression, apathy, together with chorea.

In the years that followed his wish for euthanasia became more pronounced, but his underlying thoughts slowly confined only to the car and the ability to drive. In conversations with him it seemed that the car became part of an obsessive-convulsive thought. We are now at a moment in time where the patient still repeatedly talks about euthanasia with his physicians, but where it has not come to the point yet that the patient actually asks for euthanasia.

Because of this deteriorating ability to express himself, we wonder if this obsessive-convulsive thought is the only part of the wish that remains, or that he is no longer able to express the underlying reasons and considerations for his request and what his suffering beholds. Thus is the wish now part of a psychiatric symptom or has he lost the ability to reason and express his thoughts, whilst these thoughts are still present or does he only remember part of his wish and know that the wish is connected to the car.

Is the patient we see now, with his psychiatric symptoms, a new person and do we have to continue the physician-patient relationship with the fact that you have to take these thoughts seriously. Or do these thoughts arise from a psychiatric disease, obsessive-compulsive disorder, and do we have to ignore these thoughts. We feel that if we ignore the thoughts, we are ignoring the patient. In somatic disease, for example, pain is a result of metastatic cancer, and we do not ignore the pain. Obsessive-compulsive thoughts are a result of HD and we should not ignore them. As the Supreme Court stated: the origin does not affect the extent to which the suffering is experienced.

The fact remains that it is difficult to determine if a patient with obsessive-compulsive thoughts or apathy still suffers. If the origin of the thoughts faded away due to dementia, or if cognitive deterioration is to such an extent that the content of the thoughts disappeared, is a patient still able to suffer? This is in line with the reasoning of The Health Council of The Netherlands that dementia alone is not enough ground for euthanasia. In literature it is also stated that patients with dementia do not actually suffer.[33] On the other hand several other studies and practical experience show that there are patients with dementia who suffer from their disease.[17;30] Furthermore if euthanasia is about anticipating future suffering, we argue that as physicians we are obligated to find answers to these questions.

In order to find an answer to these questions, we have to return to the translated will of the patient, look at others ways of communicating with the patient and try to find out if the patient is still able to tell his experiences with the disease and for example still able to talk about his obsessive-compulsive thoughts and what lies behind these thoughts. We described part of the answers to these questions in chapter 6, where we conclude that the basis for finding these kind of answers is a longstanding physician-patient relationship.

## Euthanasia, psychiatry and Huntington's Disease

The RRCs conclude that when evaluating a request for euthanasia in case of a psychiatric disease a physician always has to ask himself if the request was well considered and voluntary. A psychiatric disease can affect the judgement of a patient considering his situation and his understanding of the disease. A decreased understanding of the disease can affect decision making capacity. Psychiatrists mention to experience difficulties with the evaluation of both the voluntariness of the request and knowing if the patient really wants to die, thus if the request is well-considered. Regarding the voluntariness, the request can be complicated by compelling influences from outside, for example the experience of being a burden or a real influence from relatives or care givers.

Secondly, the wish to die must be a result of a process of considerations resulting in only one possible solution, death, and not the result of a sudden ebullition. A request from a patient suffering from a psychiatric disease can also be complicated by compelling pathological influences from inside. A request can only be well considered if the patient is 'free' to make his own decisions, not influenced by thoughts or 'assignments'. The key question remains if the patient put proper weight on all the relevant facts and circumstances over a longer period of time and can denominate all of these relevant facts and circumstances in light of his own illness.[34]

Furthermore a psychiatrist should pay extra attention to the unbearable suffering, which must be understandable to a certain point. The Dutch Association of Psychiatry (NVVP) advises psychiatrists to take the knowledge about the personality, medical history and biography of the patient into account. The NVVP states that unbearable suffering is partly defined by the permanency of the suffering.[34] Although from another perspective it must be emphasised that physicians must separate their own beliefs and biases from an objective capacity assessment process.[35] The NVVP has a guideline to help psychiatrists with the evaluation of an euthanasia request from patients with a psychiatric disease.[34]

The RRC advises to consult a psychiatrist is such cases, besides an independent second physician. Special attention should be paid to depressive moods. Suffering from any disease can cause a depressed mood, but this does not always mean the patient is depressed. A depressed mood and depression can influence the perception of suffering. An independent psychiatrist can evaluate if the request is influenced by a depression and thus influences the well-considered and voluntary request, or if the patient has a depressed mood, but this does not affect the request.(1) In HD psychiatric symptoms include more than just depression, which is mainly the focus of the RRCs in their annual reports.

Several points of the NVVP guideline are relevant for HD patients and should be evaluated, although not all points are always applicable. For example when a patient shows signs of apathy and anxiety, it requires a different approach to the patient with a request for euthanasia. Anxiety can behold both anxiety for future suffering and dependence as well as more existential anxiety for the future. Both can influence the request for euthanasia and should be investigated and evaluated with the patient to search for possible relief other than euthanasia, following the requirements of due care. Furthermore, for example in our case, obsessive-convulsive thoughts are part of the symptoms and signs of HD. Again the physician should investigate if the request is still well considered and voluntary.

Unbearable suffering in patients with psychiatric diseases can be difficult to determine. Not only because of the previously mentioned reasons, but also because psychiatric diseases can fluctuate in time and it is possible that spontaneous improvement occurs. This is not to be expected in HD, since psychiatric symptoms are part of a progressive disease. Furthermore, looking at the requirement of a reasonable alternative, it is questionable if a psychiatric treatment in case of psychiatric symptoms in a HD patient will lead to an acceptable improvement, in light of the just mentioned progression. After the ruling in the Chabot case, the Supreme court added an extra requirement in case of a request for euthanasia and psychiatry, namely an independent consultation of a psychiatrist, besides an independent second consultation of a physician. In case of a request from a HD patient, this will depend on the individual assessment of a patient. Do somatic and cognitive signs and symptoms prevail or mainly psychiatric? The answer to this question will determine which independent physician will be asked and if several other consultations will be held, such as by a psychiatrist or geriatrist.

## Future perspectives

HD is a disease that determines the course of life. Most patients know what their future will look like. We suspected that because of the possibilities in The Netherlands to talk about the end of life and to have a say in the end of life, many patients would think about just that. But as one physician stated during the interview, end of life wishes have always been present. Patients are just more open about them now, than they were earlier, when euthanasia and PAS were not legal yet.

As mentioned in several chapters, knowledge about the possibilities of euthanasia and/or PAS in HD and the use of advance directives is sometimes limited. In light of our exploration of the themes of the applicability of advance directives and the specific signs and symptoms of HD, we can come to some general or even explicit suggestions on how to deal with a request for euthanasia. For example should we document all wishes centrally, in order to be able as a physician to translate past wishes into the present situation? But that does not solve the problem of communication and the possibility to explore the present suffering or to determine if all possible treatments have been proposed and tried. It also does not help to determine if the wish is consistent.

To deal with a request for euthanasia or PAS in HD no definitive solution can be provided. For a patient to anticipate the future and make preparations for the future we, as physicians, can only act within the limits set by the law, although we are in a transition towards searching the boundaries of the law and placing our actions in an ethical perspective. In order to do this within the law, we have to make sure that every physician knows and more importantly understands the possibilities provided for in the law. Parliament provided possibilities when Parliament stated in the law that advance directives can be valid if the other requirements of due care are met. A physician can afford a certain amount of freedom, when the wishes are documented well, repeatedly discussed and consistent.

Our finding that HD patients in The Netherlands do think about their future and have wishes for the end of life, and because of their sometimes extensive knowledge about the future, we think that end of life wishes are also present in patients in other countries. Even though our research could not prove this assumption. The rate of suicide in HD is higher than in the general population. Is this only because of the psychiatric signs and symptoms of the disease or because patients want a say in their future, a say in their end of life?

We suggest further research should be done to investigate the wishes of patients in other countries; to investigate if end of life wishes exist. Furthermore with this knowledge it can be possible to educate treating physicians. Because at the moment a cure is not to be expected in the near future and thus guiding and treating your patients may also be providing quality of life and quality of death.

# REFERENCES

(1) http://www.euthanasiecommissie.nl. 2014.

(2) Griffiths J, Weyers H, Adams M. Euthanasia and Law in Europe. Oxford and Portland, Oregon: Hart publishing; 2008.

(3) Rechtbank Leeuwarden 21 februari 1973. (Postma case) Nederlandse Jurisprudentie 1973;183.

(4) Hoge Raad 27 november 1984. (Schoonheim case) Nederlandse Jurisprudentie 1985; 106.

(5) Hoge Raad 24 december 2002. (Brongersma case) Nederlandse Jurisprudentie 2003; 167.

(6) Engberts DP. [Autonomie op de achtergrond. Over motieven en ontwikkelingen in het euthanasiedebat]. Hoorn: 2010.

(7) Schwartz KD, Lutfiyya ZM. “In pain waiting to die”: everyday understandings of suffering. Palliat Support Care 2012 Mar;10(1):27-36.

(8) Maessen M, Veldink JH, van den Berg LH, Schouten HJ, van der WG, Onwuteaka-Philipsen BD. Requests for euthanasia: origin of suffering in ALS, heart failure, and cancer patients. J Neurol 2010 Jul;257(7):1192-8.

(9) Finlay I. Dying and choosing. Lancet 2009 May 30;373(9678):1840-1.

(10) Dupuis HM. Professional autonomy: A stumbling block for good medical practice. An analysis and interpretation. Theor Med Bioeth 2000 Sep;21(5):493-502.

(11) Jochemsen H, Ten Have H. The autonomy of the health professional: An introduction. Theor Med Bioeth 2000 Sep;21(5):405-8.

(12) Gastmans C, De Lepeleire J. Living to the bitter end? A personalist approach to euthanasia in persons with severe dementia. Bioethics 2010 Feb;24(2):78-86.

(13) Hardwig J. Is there a duty to die? Hastings Cent Rep 1997 Mar;27(2):34-42.

(14) [Termination of life on request and assisted suicide act.] (2002).

(15) ter Kuile B. Nederlandse Euthanasie-wetgeving. Een juridische beschouwing. Rotterdam: Kluwer; 2002.

(16) Wet toetsing levensbeeindiging op verzoek. Memorie van Toelichting, 26 691, no 3, Tweede Kamer der Staten Generaal, (1998).

(17) de Boer ME, Droes RM, Jonker C, Eefsting JA, Hertogh CM. [The lived-experiences of early-stage dementia and the feared suffering: an explorative survey]. Tijdschr Gerontol Geriatr 2010 Oct;41(5):194-203.

(18) Hertogh CM, de Boer ME, Droes RM, Eefsting JA. Would we rather lose our life than lose our self? Lessons from the Dutch debate on euthanasia for patients with dementia. Am J Bioeth 2007 Apr;7(4):48-56.

(19) Hertogh CM. The role of advance euthanasia directives as an aid to communication and shared decision-making in dementia. J Med Ethics 2009 Feb;35(2):100-3.

(20) www.knmg.nl/publicaties. 2014.

(21) [Early conversations about end-of-life wishes]; http://knmg.artsennet.nl/Diensten/knmgpublicaties/KNMGpublicatie/Handreiking-Tijdig-praten-over-het-overlijden-2011.htm. 2011.

(22) Bindler L, Travers D, Millet B. [Suicide in Huntington's disease: a review]. Rev Med Suisse 2009 Mar 18;5(195):646-8.

(23) Bird TD. Outrageous fortune: the risk of suicide in genetic testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1289-92.

(24) Almqvist EW, Bloch M, Brinkman R, Craufurd D, Hayden MR. A worldwide assessment of the frequency of suicide, suicide attempts, or psychiatric hospitalization after predictive testing for Huntington disease. Am J Hum Genet 1999 May;64(5):1293-304.

(25) Farrer LA. Suicide and attempted suicide in Huntington disease: implications for preclinical testing of persons at risk. Am J Med Genet 1986 Jun;24(2):305-11.

(26) Paulsen JS, Hoth KF, Nehl C, Stierman L. Critical periods of suicide risk in Huntington's disease. Am J Psychiatry 2005 Apr;162(4):725-31.

(27) Davis JK. The concept of precedent autonomy. Bioethics 2002 Apr;16(2):114-33.

(28) Beauchamp T, Childress J. Principles of Biomedical Ethics. 7 ed. New York: Oxford University Press; 2012.

(29) Sjostrand M, Helgesson G, Eriksson S, Juth N. Autonomy-based arguments against physician-assisted suicide and euthanasia: a critique. Med Health Care Philos 2013 May;16(2):225-30.

(30) de Boer ME, Hertogh CM, Droes RM, Riphagen II, Jonker C, Eefsting JA. Suffering from dementia - the patient's perspective: a review of the literature. Int Psychogeriatr 2007 Dec;19(6):1021-39.

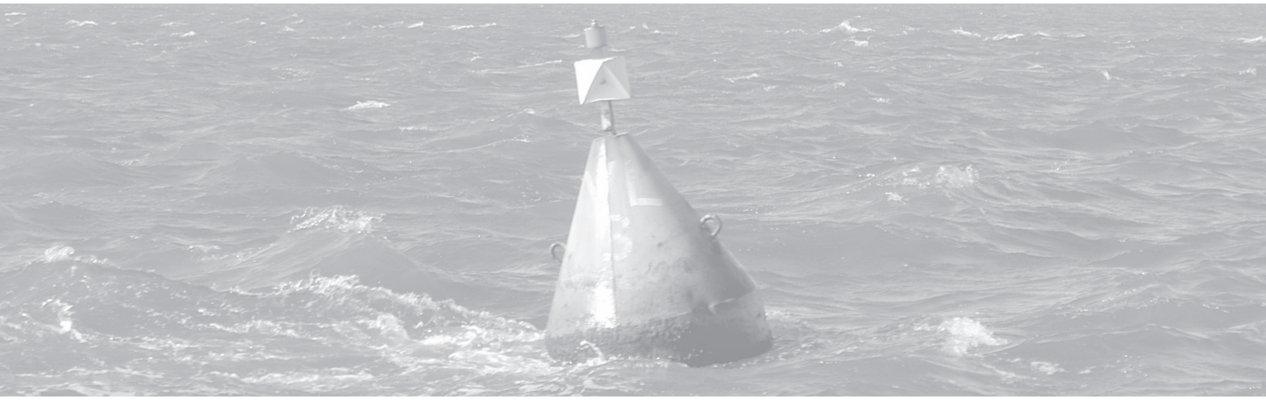
(31) van Duijn E, Kingma EM, Timman R, Zitman FG, Tibben A, Roos RA, et al. Cross-sectional study on prevalences of psychiatric disorders in mutation carriers of Huntington's disease compared with mutation-negative first-degree relatives. J Clin Psychiatry 2008 Nov;69(11):1804-10.

(32) Hoge Raad 21 juni 1994. (Chabot case) Nederlandse Jurisprudentie 1994;656.

(33) Pans E. De normatieve grondslagen van het Nederlandse euthanasierecht. Nijmegen: Wolf Legal Publishers; 2006.

(34) www.nvvp.net; euthanasia and physician assisted suicide in psychiatric patients. 2014.

(35) Moberg PJ, Rick JH. Decision-making capacity and competency in the elderly: a clinical and neuropsychological perspective. NeuroRehabilitation 2008;23(5):403-13.



# Summary

# Nederlandse samenvatting

# SUMMARY

This thesis reports on a study on wishes for the end of life, more specifically euthanasia and physician-assisted suicide, in patients with Huntington's Disease (HD) and known gene carriers.

In the introduction we report upon the development of the practice of euthanasia and physician-assisted suicide (PAS) in The Netherlands and upon the development of similar legislation in other countries (**chapter 1**). In The Netherlands the possibilities resulted in an increase in reported cases of euthanasia and PAS since 2002. Furthermore an increase of attention for this topic, especially for patients with dementia in advanced stages, resulted in the establishment of End of Life clinics.

We explored the possibilities of euthanasia and physician-assisted suicide (PAS) in HD based on the Dutch Act "Termination of Life on Request and Assisted Suicide" from 2002 (**chapter 2**). Since HD is a neurodegenerative disease, characterized by cognitive deficits leading to dementia, asking for euthanasia and PAS can be a problem for patients, because of the nature of the disease, the nature of suffering and the problem with the requirement that the request must be well-considered and voluntary. On the other hand following the wish of a patient can be a problem for physicians as well, because of the afore mentioned problems. Exploring these issues and performing a literature study showed that euthanasia and PAS are possible in patients with HD within the current boundaries of the law of 2002 in The Netherlands.

To explore if patients actually have wishes for the end of life, we interviewed patients and known gene-carriers for HD about their wishes and to study if participants are willing to talk about this topic (**chapter 3**). We discovered that many interviewed persons already had wishes for the end of life. Some of them had already talked about their wishes with family members or a physician, for others wishes concerned more of a feeling or a vague wish of what they wanted or wanted to avoid. Although our questions were not directed towards euthanasia and PAS specifically, most participants talked about the end of life and death spontaneously. The reason for thinking about the end of life and wanting to have a say in these stages of life and disease was in most cases the example of an affected parent. Not all participants were aware of the fact that a wish for euthanasia or PAS and an advance directive should be discussed with a physician repeatedly. Some attributed importance to family members and their consent, which is not a requirement of the law. Furthermore the scope of an advance directive was somewhat overestimated and the

requirement of sound professional support during the whole process of disease in light of the fulfillment of wishes for the end of life was underestimated.

After our qualitative study in Chapter 3, describing important themes which are important for HD patients when thinking about the end of life, we further investigated the wishes for the end of life of patients quantitatively (**chapter 4**). The aim of this study was to find out if there were any demographic or disease characteristics which associated with the presence of wishes and if the content of wishes was associated with any of these characteristics. For this purpose we developed a questionnaire which was sent out to all known HD patients and gene carriers registered in the Registry database in the Leiden University Medical Center. We received 134 filled out questionnaires. Seventy-five percent of respondents answered that they have some kind of wishes for the end of life. Eighty-six out of 134 responded to have thoughts about euthanasia and/or PAS. The presence of thoughts or wishes for the end of life was significantly related to being familiar with HD in the family, but not related to any other demographic or disease characteristic. Respondents with thoughts particularly directed towards euthanasia and/or PAS were of higher education and in the earlier stages of the disease. These results show that it is important for every physician to be aware of the fact that HD patients think about the end of life and these thoughts and wishes should be discussed.

After these surveys on the thoughts, wishes and opinions of HD patients and known gene carriers, we also asked physicians about their role and function when talking and thinking about the end of life. In **Chapter 5 and 6** we describe the results of our qualitative study amongst physicians, familiar with HD and their thoughts and experiences with wishes for the end of life from patients, especially euthanasia and PAS. Using a qualitative method we wished to identify themes that are important for a physician when talking about (wishes for) the end of life with HD patients. In **chapter 5** we argued that a physician has a legal, professional and moral responsibility to talk about the end of life and wishes for the end of life with patients. A physician has to play an active role in this process and especially the advance directive should have a more central role in the communication between physician and patient. In order to achieve a more active role of physicians towards the subject of advance care planning and advance directives, we proposed the use of a letter of intent. Using this letter promotes active participation of a physician in the process. Especially in the case of HD, where as mentioned earlier, cognitive decline is a key symptom of the disease, active participation from a physician, using a letter of intent as the basis of communication, can make the process of thinking and fulfilling wishes for the end of life, in light of a neurodegenerative disease, easier.

In **chapter 6** we further studied the evaluation of a request for euthanasia and/or PAS from the point of view of a physician. We identified three patterns of evaluation of a request, which varied according to the specialty of a physician. These patterns were based on different evaluation and dealing with cognitive decline, psychiatric signs and symptoms of the disease and on how to cope with response shift. The evaluation and handling of these themes in light of advance care planning was based upon a good and longstanding physician-patient relationship. Looking at the requirements of the Dutch law of 2002, the requirement of suffering came forward as a requirement that was judged upon differently, depending upon the specialty of each physician. Looking more closely at these themes, loss of decision making capacities, psychiatric signs and symptoms and response shift are themes that every physician has to cope with when talking about advance care planning with a patient. This does not only apply to The Netherlands, where euthanasia and PAS are legal, but apply to all physicians when decisions for the end of life must be made. A firm and possibly longstanding physician-patient relationship with knowledge of the patients past, present and future values, goals and wishes is of importance for all physicians.

To support our conclusions of chapter 6 we extended our study towards physicians in other countries in Europe. To find out whether these physicians also receive questions and wishes for the end of life and if advance care planning is part of their treatment and guidance of HD patients, we sent out a questionnaire to more than 500 physicians in Europe, all participants in the European Huntington's Disease Network. The results of this study are described in **chapter 7**. Unfortunately we only received a response rate of around 10%, which is not enough to draw any conclusion. Nevertheless, half of the respondents answered that their HD patients express wishes for the end of life. Opinions, upon the options for patients to have a say in their end of life, varied amongst the respondents. Some respondents indicated that the law in their country provided enough options for palliative care, or their personal opinion ensured that enough options to take care of their patients are available. Others responded that they would like to see options expanded, because wishes exist and patients seek other ways to find a solution for their wishes, for example suicide. Even though we could not draw any conclusions from this study, we can conclude that wishes for the end of life exist in HD patients in other European countries as well, even when euthanasia and/or PAS are against the law.

From this finding we draw the conclusion that physicians should prepare themselves for conversation of this nature with their patients in the near future.

The main results of our study are summarized in **chapter 8**. Thereafter, several legal and methodological issues with the Dutch “Law on Termination of Life on Request and Assisted Suicide” of 2002 are discussed, especially the difficulty with the second article, paragraph 2, stating that advance directives are ‘accordingly applicable’ in a request for euthanasia and/or PAS when all other requirements of the law are followed. In The Netherlands currently there is a debate in the media, in politics and in the general public opinion about the possibilities of euthanasia and/or PAS in case of a patient suffering from dementia or suffering from a psychiatric disease. Recent rulings of the Review Committees of Euthanasia are discussed, especially when they concern HD, and the theme of psychiatric symptoms and signs is further elaborated.

Based on this study we conclude that euthanasia and PAS are possible in case of a patient suffering from Huntington’s Disease, also based on an advance directive. Requirements in order to make this possible are a sound and possibly longstanding physician-patient relationship. Secondly a thorough knowledge of the requirements of due care and the applicability of advance directives is necessary, for patients as well as physicians. Physicians and patients should both be educated upon the possibilities within the law.

# NEDERLANDSE SAMENVATTING

Dit proefschrift beschrijft de resultaten van een studie naar levenseindewensen, meer specifiek euthanasie en hulp bij zelfdoding, bij bekende gendragers en patiënten met de ziekte van Huntington (HD).

In de Introductie beschrijven we de ontwikkeling in regelgeving en de praktijk van euthanasie en hulp bij zelfdoding in Nederland en beschrijven we de ontwikkeling van soortgelijke wetgeving in andere landen (**Hoofdstuk 1**). De mogelijkheden die de wet bood op het gebied van euthanasie en hulp bij zelfdoding resulteerden in een toename van de rapportage van verleende euthanasie en hulp bij zelfdoding vanaf 2002. Daarnaast zorgde een toegenomen aandacht voor dit onderwerp, in het bijzonder bij patiënten met dementie in een gevorderd stadium, voor de oprichting van de Levenseinde-klinieken.

We verkenden de mogelijkheden van euthanasie en hulp bij zelfdoding bij de ziekte van Huntington gebaseerd op de Wet Toetsing Levensbeëindiging op Verzoek en Hulp bij Zelfdoding uit 2002 in **Hoofdstuk 2**. Omdat de ziekte van Huntington een neurodegeneratieve ziekte is, gekenmerkt door cognitieve stoornissen die tot dementie leiden, kan vragen naar levensbeëindiging door euthanasie of hulp bij zelfdoding een probleem zijn. Dit vanwege de aard van de ziekte, de aard van het lijden en het probleem dat een verzoek om levensbeëindiging weloverwogen en vrijwillig moet worden gedaan. Anderzijds kan het voldoen aan de wens van een patiënt ook voor een arts een probleem zijn, vanwege de zojuist geschetste problemen. Na bestudering van de literatuur en het exploreren van de problemen komen we tot de conclusie dat euthanasie en hulp bij zelfdoding bij patiënten met de ziekte van Huntington in Nederland mogelijk zijn binnen de grenzen van de wet uit 2002.

Om te verkennen of patiënten ook daadwerkelijk wensen hebben voor het levenseinde, hebben we veertien patiënten en bekende gendragers voor de ziekte van Huntington geïnterviewd en gevraagd naar hun wensen. Daarnaast konden we met deze interviews onderzoeken of patiënten bereid zijn over dit onderwerp te spreken (**Hoofdstuk 3**). Uit de interviews konden we opmaken dat veel van de geïnterviewden al hadden nagedacht over het levenseinde. Enkele van hen hadden reeds met familie of een arts over hun wensen gesproken. Voor anderen bestonden de wensen meer uit een gevoel of nog niet expliciete gedachten of een latente wens over wat ze wilden of juist wilden vermijden. Hoewel onze vragen niet specifiek gericht waren op euthanasie en hulp bij zelfdoding, spraken de meeste deelnemers uit zichzelf over het levenseinde en de dood. Voor de meeste deelnemers was het voorbeeld van een aangedane ouder de reden om over het einde van het leven na te denken. Niet alle deelnemers waren zich bewust van het feit dat een wens voor euthanasie of hulp bij zelfdoding en een wilsverklaring herhaaldelijk met een arts moeten worden besproken. Sommigen gaven aan dat familieleden belangrijk zijn en dat zij moeten instemmen met de wensen, terwijl dit geen vereiste is volgens de wet. Daarnaast werd de reikwijdte van een wilsverklaring overschat en de centrale rol van de arts, in het proces van het uiten van wensen en met name het tegemoet komen aan de levenseindewensen, onderschat.

Vervolgens hebben we de wensen voor het levenseinde van patiënten kwalitatief onderzocht (**Hoofdstuk 4**). Het doel van dit onderzoek was om uit te zoeken of er demografische of patiëntgebonden karakteristieken zijn die geassocieerd zijn met de aanwezigheid van wensen. Om dit te onderzoeken hebben we een vragenlijst ontwikkeld en verstuurd naar 242 bekende HD-patiënten en gendragers die geregistreerd staan in de Registry database van het Leids Universitair Medisch Centrum. We ontvingen 134 ingevulde vragenlijsten retour. Vijfenzeventig procent (n=101) van de respondenten gaven aan dat zij wensen hadden met betrekking tot het levenseinde in enige vorm. Vierenzestig procent (n=86) gaf aan dat hun gedachten ook uitgaan naar euthanasie of hulp bij zelfdoding. De aanwezigheid van wensen of gedachten voor het levenseinde was significant geassocieerd met het feit dat de respondent bekend was met de ziekte van Huntington omdat familieleden de ziekte hebben of hadden, maar niet geassocieerd met enig ander kenmerk (demografisch of ziektegerelateerd). Respondenten met gedachten of wensen die expliciet gericht waren op euthanasie of hulp bij zelfdoding waren hoger opgeleid en in de vroegere fasen van de ziekte. Deze resultaten laten zien dat van te voren niet te bepalen is wie wensen heeft en dat het dus belangrijk is voor een arts om te vragen naar wensen en deze te bespreken.

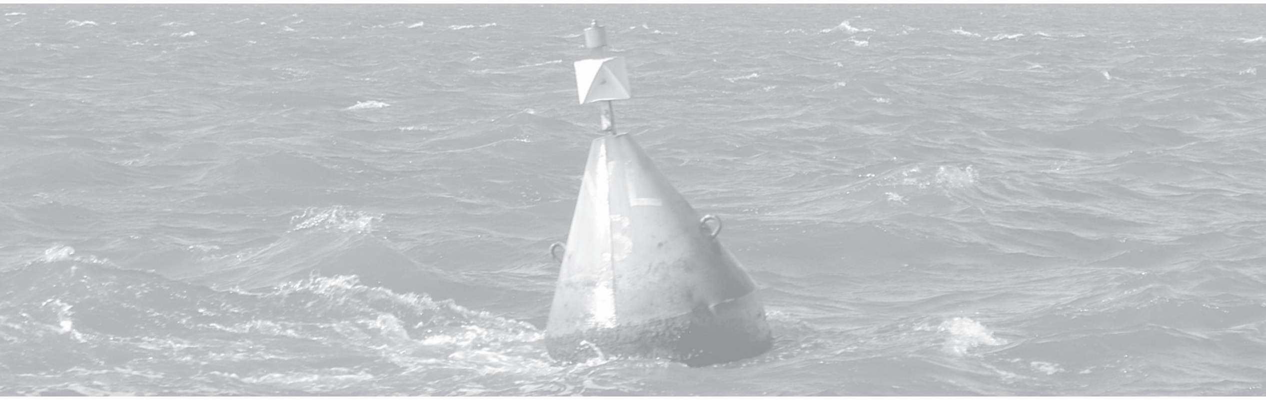
Na onze onderzoeken onder patiënten en bekende gendragers hebben we ook artsen gevraagd naar hun rol en functie wanneer ze met patiënten nadenken en spreken over het levenseinde. **Hoofdstukken 5 en 6** beschrijven de resultaten van ons kwalitatief onderzoek onder artsen, die bekend zijn met de ziekte van Huntington, over hun gedachten en ervaringen met levenseindewensen van hun patiënten, specifiek met betrekking tot euthanasie en hulp bij zelfdoding. Door te kiezen voor een kwalitatieve onderzoeksmethode wilden we thema’s identificeren die belangrijk zijn voor een arts wanneer hij spreekt over (wensen voor) het levenseinde met HD-patiënten. In **Hoofdstuk 5** beargumenteren we dat een arts een juridische, professionele en morele verantwoordelijkheid heeft om over het levenseinde te praten met patiënten. Een arts moet een actieve rol spelen in dit proces en met name de wilsverklaring zou een meer centrale rol moeten hebben in de communicatie tussen arts en patiënt. Om deze actieve rol te benadrukken en ondersteunen stellen wij voor dat arts en patiënt een intentieverklaring opstellen. Zeker in het geval van de ziekte van Huntington, waarbij zoals eerder gezegd cognitieve achteruitgang een kernsymptoom is, kan actieve participatie van de arts, met de intentieverklaring als startpunt voor de communicatie, het proces van denken over, invulling geven aan en uitvoeren van de levenseindewensen, in het kader van een neurodegeneratieve ziekte, gemakkelijker maken.

In **Hoofdstuk 6** hebben we de beoordeling en evaluatie van een verzoek voor euthanasie of hulp bij zelfdoding verder bestudeerd vanuit het gezichtspunt van de arts. We konden drie patronen identificeren wanneer een verzoek wordt beoordeeld, die varieerden al naar gelang de specialisatie van de arts. Deze patronen zijn gebaseerd op een verschillende evaluatie van de cognitieve achteruitgang, psychiatrische symptomen van de ziekte en hoe om te gaan met 'response shift'. De evaluatie en beoordeling van deze patronen in het licht van het maken van keuzen over het levenseinde was gebaseerd op een goede en langdurige arts-patiënt-relatie. Wanneer we kijken naar de vereisten van de Wet Toetsing Levensbeëindiging en Hulp bij Zelfdoding uit 2002 zien we dat het vereiste van ondraaglijk lijden naar voren kwam als een vereiste dat verschillend wordt geïnterpreteerd, afhankelijk van het specialisme van de arts. Kijkend in meer detail naar deze thema's zien we dat het verlies van wilsbekwaamheid, psychiatrische symptomen en 'response shift' onderwerpen zijn waarmee elke arts te maken krijgt als hij met HD-patiënten praat over hun wensen voor het levenseinde. Dit geldt niet alleen voor de Nederlandse situatie, maar voor alle artsen van HD-patiënten wanneer over het levenseinde gedacht en gesproken wordt en beslissingen voor het einde van het leven genomen moeten worden.

Om onze conclusies uit Hoofdstuk 6 te ondersteunen hebben we ons onderzoek uitgebreid naar artsen in andere landen van Europa. Om uit te vinden of deze artsen ook vragen krijgen over het levenseinde en/of nadenken over en beslissingen nemen over wensen voor het levenseinde onderdeel is van hun behandeling en begeleiding van patiënten met de ziekte van Huntington, hebben we vragenlijsten gestuurd naar meer dan 500 artsen die deelnemen aan het European Huntington's Disease Network (EHDN). De resultaten van dit onderzoek worden beschreven in **Hoofdstuk 7**. Helaas is slechts ongeveer 10% van de vragenlijsten geretourneerd en dit is niet voldoende om conclusies uit te trekken. Desondanks gaf de helft van de respondenten aan dat hun patiënten met de ziekte van Huntington levenseindewensen uiten. Meningen over de mogelijkheden die een patiënt heeft om zeggenschap te hebben over zijn levenseinde, varieerden. Sommige respondenten gaven aan dat de juridische mogelijkheden in hun land voldoende opties boden voor palliatieve zorg of hun persoonlijke benadering zorgde ervoor dat er voldoende opties waren om voor hun patiënten te zorgen. Anderen antwoordden dat zij graag zouden zien dat de mogelijkheden verruimd zouden worden, omdat de wensen nu eenmaal aanwezig zijn en patiënten andere wegen zoeken om hun wens in vervulling te laten gaan, bijvoorbeeld door suïcide. Ondanks het feit dat we geen harde conclusies konden trekken uit dit onderzoek, kunnen we wel stellen dat levenseindewensen ook aanwezig zijn bij patiënten met de ziekte van Huntington in andere Europese landen, zelfs wanneer euthanasie en hulp bij zelfdoding niet toegestaan zijn.

De belangrijkste bevindingen van ons onderzoek zijn samengevat in **Hoofdstuk 8**. Daarna worden enkele methodologische aandachtspunten van de Wet Toetsing Levensbeëindiging op verzoek en Hulp bij Zelfdoding uit 2002 besproken, met name de moeilijkheden met het tweede artikel, lid 2, waarin wordt gesteld dat schriftelijke wilsverklaringen van overeenkomstige toepassing zijn bij een verzoek om euthanasie of hulp bij zelfdoding wanneer alle andere vereisten van de wet worden gevolgd. Op dit moment is er in Nederland een debat gaande in de media, politiek en in de samenleving over de mogelijkheden van euthanasie of hulp bij zelfdoding wanneer een patiënt dementeert of een psychiatrische ziekte heeft. Recente uitspraken van de Regionale Toetsingscommissies Euthanasie worden besproken, met name wanneer zij over patiënten met de ziekte van Huntington gaan, en het thema van psychiatrische symptomen wordt in dit hoofdstuk verder uitgewerkt.

Gebaseerd op dit onderzoek concluderen wij dat euthanasie en hulp bij zelfdoding in Nederland binnen de kaders van de huidige wet- en regelgeving mogelijk zijn bij patiënten met de ziekte van Huntington, ook op basis van een schriftelijke wilsverklaring. Een vereiste om dit mogelijk te maken zijn een goede en indien mogelijk langer bestaande arts-patiënt-relatie. Daarnaast is een grondige kennis van de vereisten van de wet en de toepasbaarheid van de wilsverklaring noodzakelijk, zowel voor artsen als voor patiënten. Zowel artsen als patiënten zouden verder geschoold en/of voorgelicht moeten worden over de mogelijkheden die de wet hun biedt.



# Appendix

# TOPIC LIST INTERVIEWS MET ARTSEN

Gedachten over euthanasie

Gedachten over euthanasie bij Huntington

Speelt het feit dat patiënten cognitief achteruit gaan een rol bij deze gedachten

Ervaringen met wilsverklaringen bij Huntington

- wanneer bespreek je dat met patiënten
- hoe bespreek je dat
- met wie bespreek je dat (patiënten/familie)

Wanneer komt het onderwerp euthanasie bij patiënten/familie naar voren

- wat zijn de omstandigheden waaronder het onderwerp naar voren komt

Ervaringen met euthanasie bij Huntington

Is er veel verschil tussen wilsverklaringen en euthanasie in de praktijk

Beperkingen in het uitvoeren van wilsverklaringen bij Huntington

Beperkingen in het uitvoeren van euthanasie bij Huntington

Conflicten met familie/patiënten

Hoe gaat het team verder met deze vragen om, indien patiënt in een instelling verblijft

Zien artsen mogelijkheden om de zorg op dit vlak te verbeteren

# TOPIC LIST INTERVIEWS MET PATIËNTEN/ DRAGERS VAN DE GENMUTATIE/ PARTNERS

Sociale omstandigheden/werk

Ervaringen in de familie

Gedachten over euthanasie

Gedachten over wilsverklaringen

- wat zijn de mogelijkheden
- wat moet daar in staan
- wanneer komen de gedachten naar voren

Gesprekken met familie/partner over wilsverklaring of euthanasie

Ervaringen met wilsverklaringen in het verleden bij familie

Aanwezigheid van wilsverklaring en inhoud hiervan

Met wie is de wilsverklaring besproken, hoe vaak

Veranderingen in de gedachten over wilsverklaringen/euthanasie in de laatste jaren

Verbeteringen van zorg of begeleiding door artsen op het gebied van euthanasie/ wilsverklaringen

# QUESTIONNAIRE REGARDING END-OF-LIFE QUESTIONS IN PATIENTS WITH HUNTINGTON'S DISEASE

This questionnaire handles the subject of end-of-life wishes in patients with Huntington's disease (HD). In this study we want to investigate if physicians receive questions regarding the end of life from their HD patients, what kind of questions and how physicians handle and cope with these questions.

The questionnaire was drawn up after qualitative research with physicians familiar with HD and the treatment of HD patients.

We ask you to fill in this questionnaire about patient's you have treated with Huntington's disease (HD) or patients with HD you have been in contact with.

Your answers will be coded and analysed confidential. No answers will be traceable to you.

Instructions on how to fill in the questionnaire:

Usually we ask you to pick one answer of the options provided. If there is a different possibility, it will be indicated. There are no correct of false answers.

When answering a multiple choice questionnaire it is not always possible to find the correct nuance in the answers given. If possible pick the answer that is closest to your own or otherwise please use the 'other' option or the last page of the questionnaire to exemplify your answers. We kindly ask you to write down the number of the question when making extra remarks on the last page.

We thank you in advance for your cooperation.

## Questions about you

1. What is your medical speciality?
    a. Neurologist
    b. General practitioner
    c. Psychiatrist
    d. Geriatrist or physician of the elderly

2. How many years do you practice as a physician?
    a. 0-5 years
    b. 5-10 years
    c. 10-20 years
    d. 20-30 years
    e. > 30 years

3. What is your gender?
    a. Male
    b. Female

4. What is your age?
    a. 20-30 years
    b. 30-40 years
    c. 40-50 years
    d. 50-60 years
    e. > 60 years

5. In what country do you practice as a physician?

6. Are you religious?
    a. Yes
    b. No

7. If Yes, what is your religion?
    a. Catholic
    b. Protestant
    c. Islamic
    d. Other, …

8. How important is religion to you?
    a. Very important
    b. Reasonably important
    c. Not important/ not unimportant
    d. Reasonably unimportant
    e. Very unimportant

## General questions about end-of-life wishes

9. Are end of life wishes concerning euthanasia or physician assisted suicide valid in your country?
    a. Yes
    b. No

10. Are end of life wishes concerning for example do not resuscitate, treatment limitations, rejection of fluid or food etc valid in your country?
    a. Yes
    b. No

11. Sometimes there is a difference between official and how things are handled semi-official or off the record, considering wishes regarding the end of life. Are you familiar with a situation where reality of handling was different to the official requirements? If yes, could you describe the situation?

12. Are advance directives concerning euthanasia or physician assisted suicide valid in your country?
    a. Yes
    b. No

13. Are advance directives concerning other wishes regarding the end of life, for example no tube feeding, no treatment with intravenous antibiotics, DNR valid in your country?
    a. Yes
    b. No

14. What kind of advance directive? (multiple answers possible)
    a. Do-not-resuscitate orders
    b. Other treatment limitations, other than DNR
    c. Wish for euthanasia
    d. Concerning a wish to live and treatment order
    e. Concerning the appointment of a representative
    f. Other, …

## General questions about HD patients

15. Approximately how many patients with HD do you treat at the moment?

16. Approximately how many patients with HD have you treated in the past?

17. What is the main cause of death for most HD patients in your country?
    a. Pneumonia or other infection
    b. Suicide
    c. After stop taking food and fluids
    d. Medical condition other than HD
    e. Other, …

18. Where do most HD patients die in your country?
    a. At home
    b. In a nursing home
    c. In the hospital
    d. Hospice

## Questions concerning end-of-life wishes in HD

19. Do HD patients or gene carriers present themselves with wishes regarding the end of life to you?
    a. Yes, continue to question 20
    b. No, continue to question 28

Please take the last patient you had with end of life wishes in mind.

20. How did you get familiar with these wishes?
    a. I asked the patient about these wishes
    b. The patient talked to me about these wishes
    c. Family members talked to me about these wishes
    d. Professional care givers talked to me about the wishes

21. What was the reason for this HD patient to have end of life wishes?
    a. After seeing a family member with the disease
    b. Fear for future suffering
    c. Signs and/or symptoms of the disease present
    d. Staying in control of future own life
    e. Other, …

22. At what time were the wishes talked about?
    a. When in the phase of being at risk, but not tested
    b. When in the phase of being a gene-carrier, without symptoms
    c. When in the early phase of the disease, with only slight symptoms
    d. When in more advanced stages of the disease
    e. Other

23. What did these wishes consist of?
    - a. Wishes regarding place to live when they become dependent
    - b. Wishes regarding treatment of infections
    - c. Wishes regarding administration of fluid and food
    - d. Wishes regarding admittance to a hospital
    - e. Wishes regarding admittance to a nursing home
    - f. Wishes regarding death, euthanasia, physician assisted suicide
    - g. Do not resuscitate
    - h. Other, …

24. Did the family have a role in the (determination of) wishes regarding the end of life?
    - a. Yes
    - b. No

25. What kind of role (please describe)

26. What is your reaction if a patient presents himself with a wish regarding the end of life? (please describe)

Response shift turned out to be a concern in our focus group. Response shift means adapting to the situation or deviating from anticipatory beliefs.

27. Do or did you encounter response shift?
    - a. No
    - b. Yes, in what way?

## Questions about advance directives concerning end-of-life wishes

28. Do HD patients or gene carriers ever present an advance directive to you?
    a. Yes
    b. No

Please take the last advance directive in mind.

29. How did you get familiar with the advance directive?
    a. I asked the patient about the presence of an advance directive
    b. The patient told me about his advance directive
    c. Family members told me about the presence of an advance directive
    d. Professional care givers told me about the presence of an advance directive

30. When was the advance directive drawn up?
    a. When in the phase of being at risk, but not tested
    b. When in the phase of being a gene-carrier, without symptoms
    c. When in the early phase of the disease, with only slight symptoms
    d. When in more advanced stages of the disease
    e. Other

31. What was the content of advance directives?
    a. Do-not-resuscitate orders
    b. Other treatment limitations, other than DNR
    c. Wish for euthanasia
    d. Concerning a wish to live and treatment order
    e. Concerning the appointment of a representative
    f. Other, …

32. Do you think it is helpful for a HD patient to draw an advance directive?
    a. Yes, why
    b. No, why not

33. What is your reaction if a patient presents himself with an advance directive consisting of a wish regarding the end of life? (please describe)

34. What happened with the advance directive and the wish described in the advance directive? (please describe)

35. Were any actions taken? If yes please describe

36. Do you encounter response shift with respect to advance directives?
    a. Yes (please answer questions 36)
    b. No (please go to question 37)

37. How do you handle response shift with regard to an advance directive? (please describe)

Huntington's disease is also characterized by cognitive deterioration.

38. Do you think an advance directive can be valid in case of HD?
    a. Yes
    b. Yes, under certain circumstances (please describe)
    c. No, (please describe why not)

39. How do you cope with an advance directives and cognitive deterioration? (please describe)

40. Wishes regarding the end of life can be an intensively discussed subject. Physicians are searching for a way to handle these subjects, opinions have to be formed. Did your opinion change regarding end-of-life wishes in HD while practicing as a doctor?
    a. Yes, in what way
    b. No

41. What is your opinion about the present options a patient has regarding end of life wishes and the possibility of having these wishes executed?

42. What would be your wish regarding end of life wishes and the possibilities for patients to choose their end of life?

Thank you for your cooperation

# VRAGENLIJST: ‘BESLISSINGEN EN WENSEN RONDOM HET LEVENSEINDE BIJ MENSEN MET DE ZIEKTE VAN HUNTINGTON OF IN DE PREMANIFESTE FASE VAN DE ZIEKTE.’

Deze vragenlijst gaat over de vragen rondom het levenseinde van patiënten met de ziekte van Huntington of in de premanifeste fase van de ziekte.

We kijken in deze studie naar vragen die kunnen spelen bij patiënten en hun verzorgers/partners en hun gedachten en eventuele weg naar het zoeken van een antwoord of verwoorden van het antwoord. Door middel van deze vragenlijst willen wij meer inzicht krijgen in de wensen van patiënten en hun verzorgers/partners als het gaat om beslissingen en wensen rondom het levenseinde.

In deze vragenlijst komen de volgende onderwerpen aan bod:

Algemene vragen over u
Sociaal functioneren en kwaliteit van leven
Gezondheidszorg
Verblijfsplaats
Wensen rondom het levenseinde

Instructies voor het invullen:
Bij de meeste vragen is het de bedoeling dat u één antwoord invult. Als er meer dan één antwoord ingevuld kan worden, staat dat aangegeven. Er zijn geen goede of foute antwoorden. De vragenlijst bestaat uit 39 vragen.

Aangezien de vragenlijst voornamelijk uit meerkeuzevragen bestaat, kan het zijn dat het antwoord dat u zou willen geven er niet precies bij staat. In dat geval kunt u het antwoord aankruisen dat het meest de werkelijkheid benaderd of de mogelijkheid `anders` aankruisen en daar het antwoord invullen.

Indien u een vraag echt niet kunt of wilt beantwoorden, kunt u de vraag open laten.

Aan het einde van de vragenlijst is ruimte voor toelichting. Wij verzoeken u vriendelijk de toelichting per vraagnummer te noteren.

Bij voorbaat dank voor uw medewerking

## Datum van invullen:

## Algemene vragen over u

1. Wat is uw geslacht?
    a. Man
    b. Vrouw

2. Wat is uw leeftijd
    a. …

3. Wat is uw burgerlijke staat?
    a. Alleenstaand
    b. Samenwonend
    c. Getrouwd
    d. Gescheiden
    e. Weduwnaar/weduwe

4. Beschouwt u zich als behorend tot een godsdienstige groepering of voelt u zich verbonden met een bepaalde levensbeschouwelijke overtuiging?
    a. Nee
    b. Ja, namelijk…
        i. Ik ben katholiek
        ii. Ik ben hervormd
        iii. Ik ben gereformeerd
        iv. Ik ben islamitisch
        v. Anders, namelijk …

5. Hoe belangrijk is uw geloof of levensovertuiging voor u?
    a. Heel belangrijk
    b. Belangrijk
    c. Niet belangrijk/ niet onbelangrijk
    d. Onbelangrijk
    e. Helemaal onbelangrijk

6. Wat is uw hoogst afgeronde opleiding?
    - a. Geen
    - b. Lagere school
    - c. Lager beroeps onderwijs (VMBO, LTS, huishoudschool)
    - d. Middelbaar algemeen onderwijs (MULO, MAVO, VMBO-t)
    - e. Middelbaar beroeps onderwijs (MBO, MTS)
    - f. Voortgezet algemeen onderwijs (HAVO, HBS, MMS, VWO)
    - g. Hoger beroeps onderwijs (HBO, HTS, sociale academie)
    - h. Universitair onderwijs
    - i. Anders, ...

7. Was u bekend met de ziekte van Huntington in uw familie toen u de ziekte kreeg?
    - a. Ja
    - b. Nee

8. Vanaf welke leeftijd was u bekend met de ziekte van Huntington in uw familie?

    ...

9. Van wie heeft u de ziekte geërfd?
    - a. Vader
    - b. Moeder
    - c. Onbekend

## Sociaal functioneren en kwaliteit van leven

10. Belemmert de ziekte u in uw sociale bezigheden
    - a. Helemaal niet
    - b. Een beetje
    - c. Nogal
    - d. Heel erg
    - e. Weet niet

11. Werkt u op dit moment?
    - a. Ja, full time, zonder aanpassingen in mijn normale werk
    - b. Ja, full time met aanpassingen van mijn normale werk
    - c. Ja, part time, zonder aanpassingen van mijn normale werk
    - d. Ja, part-time met aanpassingen in mijn normale werk
    - e. Ja, ik doe vrijwilligerswerk
    - f. Nee, ik ben ontslagen en zit in de WW
    - g. Nee, dat lukt mij niet meer, ik zit in de WAO/WIA/ziektewet
    - h. Nee, ik heb nooit gewerkt.

12. Hoe schat u uw kwaliteit van leven in de afgelopen periode?

1 2 3 4 5 6 7 8 9
Erg slecht Neutraal Uitstekend
Weet niet

## Gezondheidszorg

13. Met welke zorgverleners heeft u tijdens uw ziekte de afgelopen periode te maken gehad?
    - a. Huisarts
    - b. Arts in verpleeghuis
    - c. Neuroloog
    - d. Andere medisch specialist
    - e. Psycholoog/maatschappelijk werker
    - f. Verzorgenden (wijkverpleging, verzorging etc)
    - g. Fysiotherapeut
    - h. Logopediste
    - i. Ergotherapeut
    - j. Diëtist
    - k. Maatschappelijk werkende
    - l. Geestelijk verzorger
    - m. Anders, namelijk ...

## Verblijfplaats

14. Waar woont u?
    a. Thuis
    b. Verzorgingshuis
    c. Verpleeghuis
    d. Ziekenhuis
    e. Anders, namelijk ...

15. Wanneer was de laatste verhuizing? (geef indien mogelijk maand en jaar aan)
    ...

16. Kunt u aangeven wat de reden was van de laatste verhuizing? (meer antwoorden mogelijk)
    a. Uit voorzorg voor eventuele latere problemen
    b. Ik had meer verzorging nodig
    c. Ik had medische behandeling nodig
    d. Het was mijn wens
    e. Het was de wens van mijn belangrijkste naasten
    f. Anders, namelijk ...

## Wensen rondom het levenseinde

17. Is de ziekte onderwerp van gesprek tussen u en uw familie/gezin?
    a. Zo ja, hoe vaak spreekt u daarover?
        i. Dagelijks
        ii. Wekelijks
        iii. Maandelijks
        iv. Ongeveer 1x per half jaar
        v. Ongeveer 1x per jaar
        vi. Minder
    b. Nee

18. Wie brengt het onderwerp meestal ter sprake?
    a. U zelf
    b. Uw partner
    c. Uw kinderen
    d. Anders, namelijk ...

19. Sprak u ooit met een van uw ouders over de ziekte, het beloop van de ziekte?
    a. Nooit
    b. Zelden
    c. Af en toe
    d. Regelmatig
    e. Vaak

20. Hebt u ooit nagedacht over wensen rondom het levenseinde? (Meerdere antwoorden mogelijk)
    a. Ja, over mogelijkheden van zorg thuis
    b. Ja, over opname in een verpleeghuis
    c. Ja, over eventueel sondevoeding
    d. Ja, over eventueel opname in een ziekenhuis als dat nodig mocht zijn
    e. Ja, over eventueel behandeling van een infectie, zoals een longontsteking
    f. Ja, over euthanasie of hulp bij zelfdoding
    g. Nee

21. Kunt u aangeven wat de reden is van het nadenken over uw wensen rondom het levenseinde? (meerdere antwoorden mogelijk)
    a. Een van mijn ouders was ziek, ik heb het meegemaakt en wil dat niet voor mezelf
    b. Angst voor de toekomst
    c. Lichamelijke aftakeling
    d. Ik wil niet afhankelijk zijn
    e. Niet meer kunnen communiceren met anderen
    f. Ik wil de controle behouden
    g. Het verlies van persoonlijke waardigheid, ontluistering

   h. Ik wil niet in een verpleeghuis opgenomen worden
   i. Ik weet dat het einde komt, dan kan ik er maar beter nu al over nadenken.

22. Zou u later sondevoeding willen?
    a. Ja
    b. Nee

23. Zou u later opgenomen willen worden in een verpleeghuis
    a. Ja
    b. Nee

24. Denkt u wel eens na over euthanasie of hulp bij zelfdoding?
    a. Ja
    b. Nee

25. Hebt u wel eens gesproken met uw partner/gezin over wensen rondom het levenseinde?
    a. Nee
    b. Ja

26. Hebt u wel eens met uw huisarts gesproken over wensen rondom het levenseinde?
    a. Ja
    b. Nee

27. Hoe omschrijft u de relatie met uw huisarts?
    a. Goed
    b. Voldoende
    c. Matig
    d. Slecht

28. Als u nog niet met uw huisarts heeft gesproken over uw wensen, wat is hiervoor de reden?
    a. Ik ben er nog niet aan toe
    b. Ik zie er tegen op, ik vind het eng om hierover te beginnen
    c. Ik stel het steeds uit, het komt er gewoon niet van.
    d. Ik heb geen wensen, ik neem het leven zoals het komt.
    e. Ik voel met bezwaard dit onderwerp met mijn huisarts te bespreken
    f. Mijn huisarts is gelovig, dus ik denk dat hij negatief zal reageren.
    g. Een eerdere reactie van mijn huisarts doet mij vermoeden dat hij negatief zal reageren
    h. Ik heb nauwelijks contact met mijn huisarts, dus ben hiervoor niet bij hem/haar langs geweest.
    i. Mijn huisarts weet erg weinig van de ziekte. Daarom ben ik zeker nog niet over mijn wensen begonnen.

29. Zijn uw wensen veranderd in de afgelopen jaren?
    a. Nee, de wensen zijn niet veranderd
    b. Ja
    c. Niet van toepassing

30. Kunt u aangeven wat de reden was van de verandering van de wensen?

31. Kunt u aangeven hoe de wensen veranderd zijn? (wat waren ze eerst en wat zijn ze nu?)

32. Hebt u een wilsverklaring?
    a. Ja (ga verder met vraag 33)
    b. Nee (dit is de laatste vraag, ga zo nodig naar de toelichting)

33. Wat is dit voor wilsverklaring?
    a. Euthanasieverzoek
    b. Behandelverbod
    c. Volmacht
    d. Niet reanimeren verklaring
    e. Levenswensverklaring
    f. Anders, namelijk ...

34. Waar is uw wilsverklaring van afkomstig?
    a. NVVE
    b. Notaris
    c. Eigen geschreven
    d. Anders

35. Bent u lid van de Nederlandse Vereniging voor vrijwillig levenseinde (NVVE)?
    a. Ja
    b. Nee

36. Hebt u deze wilsverklaring recent besproken met uw partner/gezin/familie?
    a. Ja
    b. Nee

37. hoe vaak bespreekt u deze verklaring met uw partner/gezin of familie?
    a. Ik heb hem alleen besproken toen ik hem maakte
    b. Ik bespreek hem ongeveer 1x per jaar
    c. Ik bespreek de verklaring ongeveer 2x per jaar
    d. Ik bespreek de verklaring vaker, namelijk

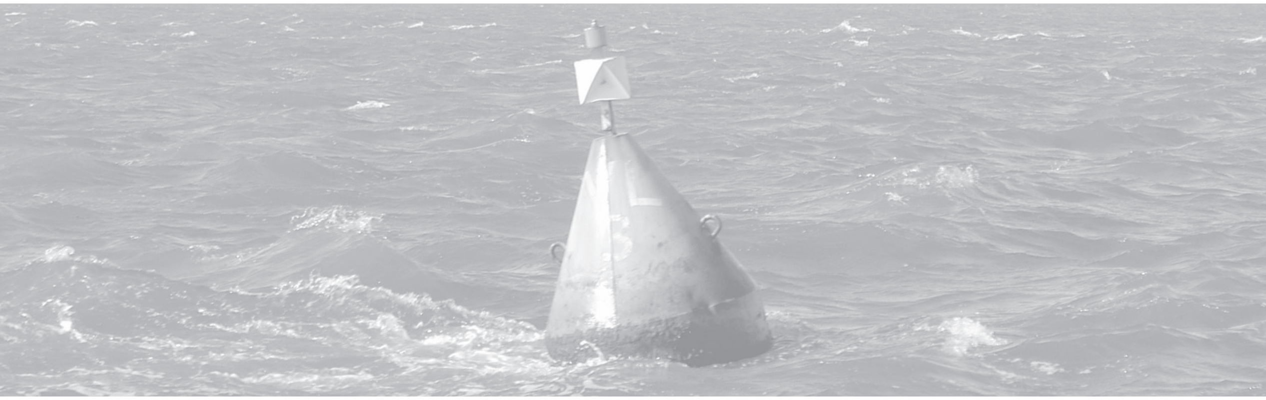
38. Hebt u deze wilsverklaring recent besproken met uw huisarts of specialist?
    a. Ja
    b. Nee

39. Hoe vaak bespreekt u deze verklaring met uw huisarts?
    a. Ik heb de verklaring alleen besproken toen ik hem maakte
    b. Ik bespreek de verklaring ongeveer 1x per jaar
    c. Ik bespreek de verklaring ongeveer 2x per jaar
    d. Ik bespreek de verklaring vaker, namelijk ...

Dank voor het invullen van de vragenlijst.
Graag verzoeken wij u de vragenlijst in de bijgestuurde envelop terug te sturen.

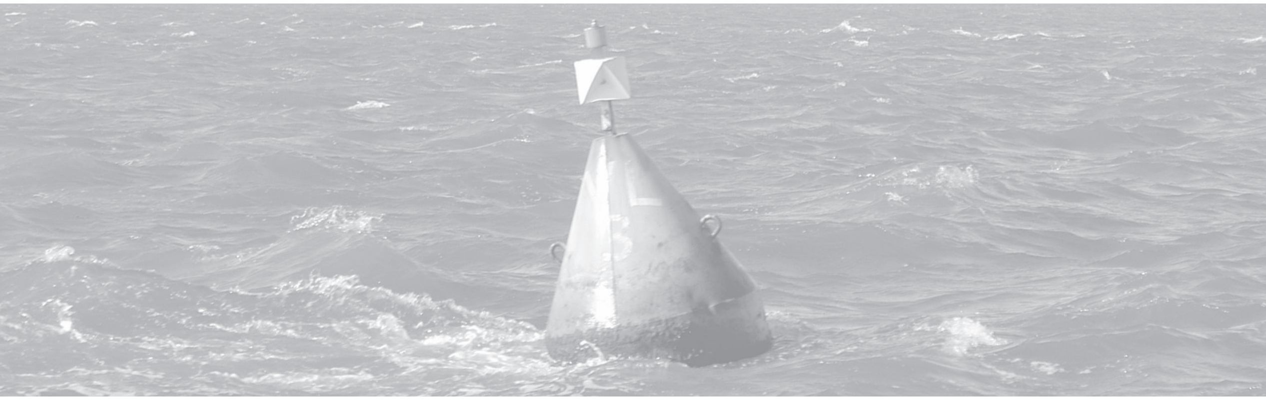
Mw mr drs S.J. Booij Prof dr R.A.C. Roos

Ruimte voor toelichting op bovenstaande antwoorden
(wilt u bij de toelichting het nummer van de vraag noteren?)



# Dankwoord

Mijn dank gaat uit naar allen die hebben meegewerkt aan de totstandkoming van dit proefschrift, in het bijzonder de patiënten die hebben toegestemd in een interview of die de tijd en energie hebben gevonden voor het beantwoorden van de vragen uit de vragenlijst over dit moeilijke, soms confronterende en emotionele onderwerp. Ook dank ik de collegae die openhartig met mij spraken over hun ervaringen, meningen, wensen en gedachten.



# Curriculum Vitae

Suzanne José Booij werd op 10 mei 1980 geboren in Delft. Na het cum laude behalen van het gymnasiumdiploma aan het Westland College te Naaldwijk in 1998, studeerde zij Geneeskunde en Nederlands Recht aan de Universiteit Leiden. Tijdens haar studie liep zij stage op de afdeling arbeidsrecht en bedrijfsrecht bij advocatenkantoor NautaDutilh te Amsterdam. De wetenschappelijke stage van de studie Geneeskunde en de afstudeerscriptie voor Nederlands Recht was een gecombineerd traject, in samenwerking met de afdeling Transplantatiechirurgie en Pathologie van het Erasmus MC te Rotterdam, afdeling Burgerlijk Recht van de Rijksuniversiteit Leiden en de sectie Ethiek en Recht van de Gezondheidszorg van het LUMC te Leiden, onder leiding van prof mr J.H. Nieuwenhuis, prof mr dr D.P. Engberts, prof dr H.W. Tilanus en drs P. Zondervan. Op 24 november 2004 behaalde zij haar doctoraalexamen in zowel het Nederlands Recht als in de Geneeskunde. Op 26 november 2006 behaalde zij haar artsenbul. Aansluitend startte zij als ANIOS Neurologie in het LUMC en per 1 juli 2007 als AIOS met prof dr R.A.C. Roos als opleider.

Tijdens haar opleiding was zij actief als bestuurslid van de LAD, de Landelijke vereniging voor Artsen in Dienstverband, en bestuurslid en voorzitter van de LVAG, de Landelijke Vereniging voor AIOS. Namens de LVAG nam zij in 2010 deel aan de Stuurgroep Evaluatie Opleidingsfonds van het ministerie van VWS.

In 2011 startte zij met haar promotieonderzoek naar wensen voor het levenseinde bij patiënten met de ziekte van Huntington, resulterend in dit proefschrift. In september 2013 rondde zij de opleiding tot neuroloog af. Zij werkt sinds 1 oktober 2013 als neuroloog in het Canisius Wilhelmina Ziekenhuis te Nijmegen.

Suzanne is getrouwd met Tijmen Augustijn en woont in Oegstgeest.