Wishes for the end of life in Huntington's Disease. Observations and reflections, initiated in The Netherlands

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Citation

Booij, S. J. (2014, November 12). *Wishes for the end of life in Huntington's Disease. Observations and reflections, initiated in The Netherlands*. Retrieved from https://hdl.handle.net/1887/29740

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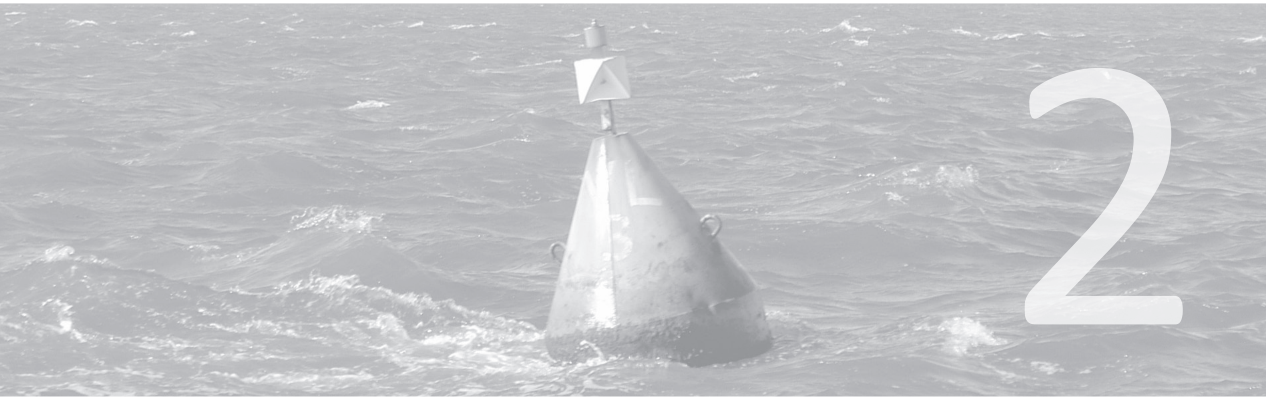
Title: Wishes for the end of life in Huntington's Disease. Observations and reflections, initiated in The Netherlands
Issue Date: 2014-11-12



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

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Published in part in International Psychogeriatrics 2013;25(2):339-40.

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.