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Title: Wishes for the end of life in Huntington’s Disease. Observations and reflections, initiated in The Netherlands
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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). 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. 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.

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.

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. 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.\textsuperscript{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.\textsuperscript{6}

**Table 1.1** The requirements of due care

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<tr>
<td>1</td>
<td>The physician is convinced that the patient’s request was voluntary and well considered</td>
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<tr>
<td>2</td>
<td>The physician is convinced that the patient’s suffering was unbearable and there was no prospect of improvement</td>
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<tr>
<td>3</td>
<td>The physician informed the patient about his situation and prospects</td>
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<tr>
<td>4</td>
<td>The physician and patient were convinced that there was no reasonable alternative in light of the patient’s situation</td>
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<td>4</td>
<td>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</td>
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<tr>
<td>5</td>
<td>The physician terminated the patient’s life or provided assistance with suicide with due medical care and attention</td>
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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.\textsuperscript{6}

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


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