Chapter 1

*General introduction*
Death has always been an unavoidable part of living. The major causes of death in developed European countries have shifted from predominantly infectious diseases towards more degenerative diseases such as cancer and cardiovascular disease\(^1\). Also, demographic changes and advances in hygiene and medicine have led to ageing of the population. As a result of these developments, sudden death has become less common. Towards the end of life, most people in developed countries acquire a serious progressive illness that increasingly interferes with their usual activities until death\(^2\). One of those progressive illnesses is Chronic Obstructive Pulmonary Disease (COPD). This thesis presents a series of studies about illness experiences of patients with advanced COPD. This introduction provides the necessary background information about palliative care, COPD, and different illness trajectories at the end of life. This chapter will end with the aim, research questions and outline of the thesis.

**Palliative care**

Palliative care originated as end-of-life care in the 1960s. At that time, it focused on providing symptom control and psychosocial and family support during the terminal phase of a life-limiting disease. Since then, its meaning and scope of practice have expanded far beyond its roots\(^3\). In the early 1980’s, the World Health Organisation (WHO) Cancer Unit began the development of a global initiative to advocate for pain relief and opioid availability at the end of life. Since then, WHO has broadened its approach to palliative care. There is now wide recognition that the principles of palliative care should be applied as early as possible in the course of any chronic, ultimately fatal illness. The physical, emotional, and spiritual needs of the patient are all considered important concerns in palliative care\(^4\). The most recent WHO definition of palliative care dates from 2002: “Palliative care is an approach that improves the quality of life of patients and their families facing the problems associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual”.
Following the same WHO definition, palliative care contains the following elements:

Palliative care...
- Provides relief from pain and other distressing symptoms
- Affirms life and regards dying as a normal process
- Intends neither to hasten or postpone death
- Integrates the psychological and spiritual aspects of patient care
- Offers a support system to help patients live as actively as possible until death
- Offers a support system to help the family cope during the patient’s illness and in their own bereavement
- Uses a team approach to address the needs of patients and their families, including bereavement counselling, if indicated
- Will enhance quality of life, and may also positively influence the course of illness
- Is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications

**Chronic Obstructive Pulmonary Disease (COPD)**

COPD is a chronic, potentially fatal progressive condition of the lungs. The American Thoracic Society (ATS) and the European Respiratory Society (ERS) maintain a slightly different definition of COPD from that of the Global Initiative for Chronic Obstructive Lung Disease (GOLD)\(^5\)\(^-\)\(^7\). GOLD has defined COPD in the following way: “COPD is a preventable and treatable disease with some significant extrapulmonary effects that may contribute to the severity in individual patients. Its pulmonary component is characterized by airflow limitation that is not fully reversible. The airflow limitation is usually progressive and associated with an abnormal inflammatory response of the lung to noxious particles or gases”\(^7\). Both this definition by GOLD, as well as the ATS definition points to the preventability and treatability of COPD, in which airflow limitation is usually progressive and not fully reversible and mention systemic or extrapulmonary elements, along with the role of exposure to noxious particles or gases\(^5\). Cigarette smoking is the most commonly encountered risk factor for COPD worldwide, although in many countries, air pollution resulting from the burning of wood and other biomass fuels has also been identified as a COPD risk factor. Once developed, COPD and its comorbidities cannot be cured and thus
must be treated continuously to reduce symptoms, improve quality of life, reduce exacerbations, and possibly reduce mortality.\(^7\)

**Epidemiology**

COPD is a serious health problem. According to the World Health Organisation, COPD ranked fourth in leading causes of death globally in 2004 (3 million deaths in total, 5.1 percent of total deaths worldwide). COPD is predicted to be the third leading cause of death globally in the year 2030.\(^8\) COPD is also associated with a substantial economic burden. Most direct costs are associated with hospitalisation (usually due to exacerbations). COPD-related costs therefore increase markedly with severity based on FEV\(_1\)\(^5,9,10\).

In the Netherlands, the prevalence of COPD based on data from general practice registers was estimated to be 176.500 men and 139.900 women (22,0 per 1.000 men and 17,1 per 1.000 women) in the year 2003.\(^11\) Based on the same general practice registers data, 17.200 men and 16.400 women are estimated to be newly diagnosed yearly (2,1 per 1.000 men and 2,0 per 1.000 women). In 2004 3.381 men and 2.281 women died from COPD (42,0 per 100.000 men and 27,7 per 100.000 women). The life expectancy of men with COPD is on average 8 years shorter than that of men without COPD (including 3 years of healthy life). Women live on average 10.5 years shorter than women without COPD (including 4 years of healthy life).\(^12\)

**Illness trajectories at the end of life**

Recent studies have described that different illness trajectories at the end of life are characterised by their own typical patterns of functional decline\(^2,13,14\). These differences between trajectories may be important for the provision of palliative care in varying populations. Besides sudden death, three distinct trajectories are identified\(^2\):

*Short period of evident decline, typically cancer (Figure 1.a.)*

This trajectory describes a reasonably predictable decline in physical health over a period of weeks, months, or years. This trajectory is mostly followed by cancer patients.
**Prolonged dwindling (Figure 1.b.)**
This trajectory is followed by people who die at an older age of either brain failure or generalized frailty of multiple body systems. This trajectory describes progressive disability from an already low baseline of cognitive or physical functioning.

**Long term limitations with intermittent serious episodes (Figure 1.c.)**
Patients who follow this trajectory are usually ill for many months or years with occasional acute, often severe, exacerbations. Each exacerbation may result in death, and although the patient usually survives many such episodes, a gradual deterioration in health and functional status is typical. This trajectory is typically followed by patients with end-stage heart failure and COPD.

**Figure 1.a. Typical illness trajectory for cancer patients**

![Typical Illness Trajectory for Cancer Patients]

**Figure 1.b. Typical illness trajectory for older frail patients**

![Typical Illness Trajectory for Older Frail Patients]
These three illness trajectories each follow their own typical pattern of functional decline. Figure 1.c. is mostly associated with the functional decline of patients in the end-stage of COPD. This illness trajectory is very different from that of cancer and frailty at the end of life. It is therefore expected that also care needs and demands of COPD patients will differ from those of cancer patients with advanced disease; the more traditional recipients of palliative care. The research described in this thesis set out to explore problems, needs and preferences of COPD patients with advanced disease.

**Figure 1.c. Typical illness trajectory for patients with end-stage heart and lung failure**

![Typical illness trajectory for patients with end-stage heart and lung failure](image)

[Figure 1.a. – 1.c.: Adapted from Murray et al. 2005²]

**COPD and palliative care**

COPD is thus acknowledged as a serious health problem with a considerable disease burden. While it is recognised that COPD patients have a shorter life expectancy than their healthy counterparts, it is still difficult to determine when exactly a COPD patient is likely to die. This is due to the highly unpredictable disease trajectory (Fig 1.c.). This trajectory is characterised by long term limitations with intermittent serious episodes. Patients are usually severely impaired for a long period of time with occasional acute exacerbations. Although these exacerbations are severe and may result in death, patients usually survive many such episodes⁵. This means that the last phase of COPD may be long and indistinct, and makes the exact timing of death problematic. Unsurprisingly, prognoses for individual patients with COPD are described to be inaccurate¹⁵,¹⁶. Therefore, the death of a COPD patient often feels sudden and unexpected for family members¹⁷. Studies have not yet provided a clear definition of the end-stage in COPD. All major international guidelines
nowadays adopt the GOLD classification of disease severity, based on spirometric values (Table 1.1).

<table>
<thead>
<tr>
<th>GOLD-stage</th>
<th>FEV&lt;sub&gt;1&lt;/sub&gt; / FVC</th>
<th>FEV&lt;sub&gt;1&lt;/sub&gt; (% of predicted value)</th>
<th>Frequency distribution in the Netherlands</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I: Mild</td>
<td>&lt; 0.70</td>
<td>&gt; 80</td>
<td>28 %</td>
</tr>
<tr>
<td>Stage II: Moderate</td>
<td>&lt; 0.70</td>
<td>50 – 80</td>
<td>54 %</td>
</tr>
<tr>
<td>Stage III: Severe</td>
<td>&lt; 0.70</td>
<td>30 – 50</td>
<td>15 %</td>
</tr>
<tr>
<td>Stage IV: Very severe</td>
<td>&lt; 0.70</td>
<td>&lt; 30 (or &lt; 50 and chronic respiratory failure)</td>
<td>3 %</td>
</tr>
</tbody>
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FEV<sub>1</sub>: forced expiratory volume in one second; FVC: forced vital capacity; respiratory failure: arterial partial pressure of oxygen (PaO<sub>2</sub>) less than 8.0 kPa (60 mm Hg) with or without arterial partial pressure of CO<sub>2</sub> (PaCO<sub>2</sub>) greater than 6.7 kPa (50 mm Hg) while breathing air at sea level

Although it is now widely recognised that the principles of palliative care should be applied as early as possible in the course of any chronic, ultimately fatal illness<sup>4</sup>, the unpredictable disease trajectory of COPD poses a challenge for the provision of high quality palliative care in this group of patients. Nonetheless, the sparse literature on end-stage COPD has shown that patients with advanced COPD may benefit from the provision of the palliative care approach<sup>20,21</sup> because they suffer from poor symptom control,<sup>22</sup> especially breathlessness,<sup>17,20,23-25</sup> anxiety<sup>17,25,26</sup> and depression<sup>17,25,27</sup> which often leads to social isolation,<sup>20</sup> increased dependency on others<sup>28</sup> and poor quality of life.<sup>29</sup> Despite the wider focus towards palliative care for other, non-malignant illnesses as described in the latest WHO definition<sup>4</sup>, palliative care has historically focused on cancer disease trajectories, and specialised services for end-stage COPD patients are still in a developing stage<sup>16,30,31</sup>. Several studies have shown that patients in the end-stage of COPD do not have similar access to specific palliative care services when compared to patients suffering from advanced lung cancer<sup>23,29,32</sup>. 
**Study aim and research questions**

In summary, COPD is a major health problem which represents a substantial economic and social disease burden, especially in advanced disease when exacerbations become frequent and severe. COPD also causes thousands of patients to die each year in the Netherlands. Still, at the same time, the provision of high quality palliative care to these patients is problematic. Little is known about the problems and demands for care in patients with end-stage COPD. This situation was the starting point of this research. The central aim of the study was to explore and better understand the problems, needs and preferences of patients with end-stage COPD. From this patient perspective, recommendations for improving palliative care provision will be drawn. The central aim of the study was operationalised by formulating the following research questions:

1. What is known about health care needs in end-stage COPD? What are specific areas where knowledge about needs is still lacking?
2. How does the quality of life of end-stage COPD patients compare to that of end-stage lung cancer patients?
3. How does the quality of life and functional status change over time in end-stage COPD patients?
4. How do patients with end-stage COPD express their health care needs?
5. To what extent can a better understanding of the end-stage in COPD be used to improve palliative care provision?

**Study design**

To answer these research questions, a mixed-methods design was adopted using both quantitative and qualitative research methods. GOLD stage IV COPD patients were identified from medical records of outpatient clinics of four participating hospitals and one centre specialising in asthma and COPD. Patients completed a self-administered questionnaire every 3 months over a period of one year or less if patients were unable to continue to participate due to increasing disease severity or death. A subgroup of patients was invited for an in-depth interview about their illness experiences. The sampling of this subgroup was done purposefully to ensure that a wide variety of patients’ experiences was recorded. A few patients were interviewed twice because the first interview did not cover all topics from the topic list, or because something changed in the patient’s
situation. Data gathered by both research methods were used as data-triangulation and were integrated in the interpretation phase of this study.

Outline of the thesis
Chapter 2 presents a structured review of the literature on health care needs in end-stage COPD patients. In this chapter an overview is given on what is already known about health care needs in end-stage COPD and in which specific areas knowledge is still lacking. Chapter 3 presents a comparison of health-related quality of life of end-stage COPD patients with more traditional recipients of palliative care: end-stage lung cancer patients. In chapter 4, results from the prospective longitudinal study are presented. This chapter will describe the changes in health-related quality of life and functional status in end-stage COPD patients during one year. Results from the qualitative part of the study are presented in chapters 5 and 6. In chapter 5, the phenomenon that end-stage COPD patients do not actively express a wish for help, despite their problems in daily life, is described and explained. Chapter 6 provides recommendations for the improvement of palliative care provision for end-stage COPD patients. In the general discussion in chapter 7, reflections on the findings and implications of the study will be given.
References


(26) Bailey PH. The dyspnea-anxiety-dyspnea cycle--COPD patients' stories of breathlessness: "It's scary /when you can't breathe". *Qual Health Res* 2004;14:760-78.


