



Key points

- ▶ Most patients with COPD die during acute exacerbations; in those who have been treated with acute NIV, almost 80% will require admission in the following year.
- ▶ A comprehensive management plan for end-stage lung disease patients needs to address symptom burden, provide information, support carers and assist with end-of-life planning.
- ▶ Management should include optimisation of treatment of airflow obstruction and pulmonary rehabilitation. In some patients, pharmacotherapy for dyspnoea, panic attacks, depression and insomnia have a key role, but the importance of practical measures, including stairlifts, bath aids and social support, should not be underestimated.
- ▶ Advance directives enable patients to give direction to the medical team about treatment preferences and should be part of routine care, especially in individuals who have already experienced an acute hypercapnic exacerbation.

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Care of end-stage lung disease

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Educational aims

- ▶ To raise awareness of the symptom load of end-stage lung disease.
- ▶ To examine trajectories of end-stage disease.
- ▶ To discuss treatment strategies.

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Summary

Individuals with end-stage lung disease have a substantial symptom burden, which is often poorly addressed compared to patients with malignant disease. Recognition of the disease trajectory in the last few years of life enables a symptom-based approach that combines palliative care with active treatment of exacerbations where appropriate. Discussion of advance directives is helpful for both the patient and the physician.

The management of end-stage lung disease has moved from the periphery to centre stage in respiratory medicine over the last few years, but approaches are often inconsistent. This is exemplified by the fact that current national, European and global chronic obstructive pulmonary disease (COPD) guidelines make few recommendations regarding end-of-life issues. Until recently, little research effort was directed towards the healthcare needs of COPD patients in their last years of life, despite the fact that as many patients with COPD die each year as those with lung cancer, and health decline occurs over a substantial period before that. Secondary (hospital) care has tended to focus on the treatment of airflow obstruction, acute exacerbations and, more recently, pulmonary rehabilitation, although provision of rehabilitation facilities does not meet requirements in many areas. Respiratory physicians are also often involved in the care of patients with respiratory insufficiency as a result of pulmonary fibrosis, or ventilatory pump failure due to muscular

dystrophies, motor neurone disease or chest wall disorders. For all these groups, a unified approach linking community, primary and secondary care is often lacking.

Disease trajectory of COPD in the last years of life

It is important to realise that the clinical course in end-stage disease varies between conditions (figure 1). For COPD patients, a gradual deterioration in airflow obstruction and activity levels, punctuated by acute exacerbations, is the norm, which differs from the more predictable decline in lung cancer patients.

In a study of COPD deaths in London (UK), ELKINGTON *et al.* [1] found that 55% of the survey group were male with an average age of 77 years. Demographic data were obtained from the person who reported the death. A remarkably high level of symptoms was reported in the 12 months before death:



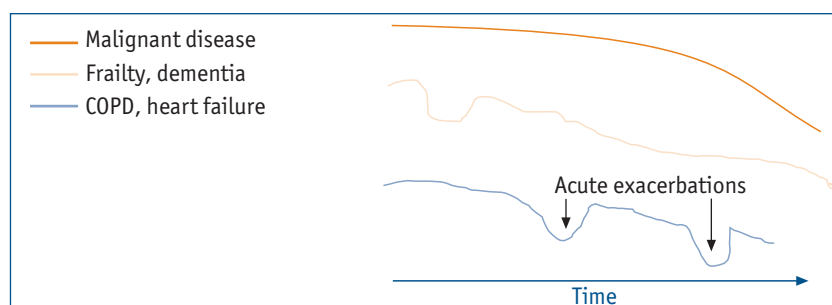


Figure 1
Predicting the natural history of end-stage conditions. The diagram shows the trajectories of some illnesses and patients' functional decline.

virtually all (98%) individuals were breathless, 77% had low mood and poor sleep quality, and 53% were subject to panic attacks. More surprisingly, pain affected 72%. Treatments offered were effective at reducing breathlessness only 57% of the time.

Importantly, less than a fifth of those with psychological symptoms had received help with these. Contact with medical services was sporadic, and only ~50% had received regular check-ups from their general practitioner. Almost 50% had experienced three or more hospital admissions in the year before death, although relatives reported that they thought hospital discharge had been premature in almost 40% of cases. This study demonstrates the major impact of COPD on healthcare resources, but it also highlights missed opportunities to help these individuals and their families. Support from social services was very limited, with 70% of families reporting that they received none. Moreover, the majority of care seemed to be directed towards intervention at the time of acute exacerbations rather than a longer-term strategy. Any such comprehensive management plan needs to address symptom burden, provide information, support carers and assist with end-of-life planning.

Understanding the prognosis

There are several major barriers to planning end-of-life care, especially in COPD patients. Prognosis may be hard to predict, since a gradual decline, punctuated by acute exacerbations, is the usual course, but it is difficult to know which exacerbation will be the last. Perhaps, as a consequence, difficult discussions and decision making may be deferred, and patients may not realise that their condition has reached a terminal phase. This makes end-stage planning problematical and may explain why relatively few patients complete advance directives. Some (40%) of the main carers of COPD patients

interviewed by ELKINGTON *et al.* [1] were not aware that the deceased was likely to die. The carers felt that ~63% of the deceased realised definitely or probably that they might die; however, a substantial number of patients were unclear about their own prognosis. Clearly, these individuals are unable to plan adequately for death if they do not know it is imminent. Regarding the place of death, it is also important to note that significantly more carers of subjects who died at home felt this was the right place to die as opposed to informants of those who died in hospital ($p=0.008$). In England, a current Department of Health initiative is aiming to improve end-of-life care by ensuring more patients die in the place of their choice (usually home).

CHU *et al.* [2] have recently shown that the prognosis is poor in individuals who receive non-invasive ventilation (NIV) for acute hypercapnic exacerbations of COPD. In this study of 110 survivors of an acute episode of ventilatory failure treated with NIV (average age 73 years), virtually 80% required re-admission, mortality rate was 49% and >63% had experienced another life-threatening event in the following year. These patients spent an average of 12% of the following year in hospital. Symptom load (Medical Research Council (MRC) dyspnoea score) was predictive of short survival. These findings are consistent with another study that suggested hypercapnic COPD patients have a worse prognosis [3]. The results of these studies enable us to target patients in whom end-of-life strategies and discussions are entirely appropriate. Other opportunities to raise this subject are during pulmonary rehabilitation sessions, although general topics are easier to address in group meetings than specific management plans. For example, HEFFNER *et al.* [4] carried out a two-site prospective evaluation of advance directive education during a pulmonary rehabilitation course to assess the effects on completion of: 1) living wills; 2) durable powers of attorney (*i.e.* identifying a proxy decision maker); 3) patient-physician discussion about end-of-life issues; 4) decisions about life support; and 5) patient impression that their physician understood their end-of-life preferences. The group that received education on these topics was subsequently significantly more likely to discuss these issues, complete advance directives and feel more assured that their physicians understood their preferences. It has been demonstrated that COPD patients in particular require further information on their disease, its likely course and treatment from their doctors, but may not necessarily raise these

topics without prompting. The way in which the topics are raised is also important. Most individuals welcome discussion, but this is usefully directed towards the symptom-control approach throughout the remainder of the patient's life, rather than an exclusive focus on their death. Striking a realistic and kindly balance between maintaining hope and a pragmatic expectation of deterioration is part of the clinical judgement healthcare workers should exercise and adapt to the individual.

What patients want to know

FRIED *et al.* [5] have examined treatment preferences regarding life-sustaining interventions in a range of patients with limited life expectancy due to cancer, congestive cardiac failure or COPD. Individuals aged ~73 years were asked whether they would wish to receive a given treatment if the outcome was certain or if there were differing likelihoods of an adverse outcome. Crucially, participants were able to balance the outcome against the burden of treatment (*e.g.* length of hospital stay, invasiveness of procedure). For example, a low-burden treatment that returned the individual to previous functioning level would be accepted by nearly all participants (98.7%), but 11.2% would not accept this option if the treatment had a high burden. Conversely, if the likely outcome was survival but with severe functional or cognitive impairment, 74.4% and 88.8% would not want to receive low- and high-burden treatment, respectively. There was no difference between choices between the diagnostic groups, although there was a trend for COPD and cancer patients to refuse high-burden therapy. This study demonstrated that patients can compute varying outcomes, and that the impact on functional and cognitive outcomes plays a greater part in preferences than in survival itself. This should help inform discussion with patients. Clearly, they also need to know the probable consequences of non-intervention to make a valid decision.

Communication and advance directives

Advance directives formulated and recorded by a competent individual enable that person to give direction to healthcare providers about their choice of treatment in particular circumstances.

These can cover treatments that individuals would wish to receive if they developed respiratory failure, whether they would wish to be resuscitated and how these decisions would change if they were, for example, senile or unlikely to regain consciousness. Over the last 5–10 years, growing numbers of people have taken up this option, although a UK survey has suggested that, so far, only 2% of the population has completed these.

Recent work has examined barriers to end-of-life discussions between the patient and the care team. In a focus group analysis of oxygen-dependent COPD patients and their physicians, KNAUFT *et al.* [6] found that only 32% of patients had taken part in such discussions with their physician. Commonly cited barriers were "I'd rather concentrate on staying alive" and "I'm not sure which doctor will be taking care of me". Physicians ranked the most important barriers as "There was too little time during our appointment





to discuss everything we should", or "I worry that discussing end-of-life care will take away his/her hope", and "The patient is not ready to talk about the care he/she wants if he/she is sick". It is evident that this topic is easier to defer on both sides. Conversely, the authors found a number of facilitators that made effective communication more likely. These included the patient's experience of friends or family who had died, the fact they trusted their physician, the feeling that their physician was good at caring for their lung disease and that he/she viewed them as person rather than focusing purely on their lung condition.

Although commonly viewed as a method of increasing autonomy and improving communication between the patient, family and healthcare team, there is some evidence that advance directives are not adhered to consistently. This may occur in circumstances that the healthcare team feel may not replicate those the patient had in mind when the advance directive was drafted [7]. Nonetheless, advance directives or living wills represent important steps forward in facilitating dialogue between the patient, family and the medical team.

Most advance directives concern decisions about treatment options that the individual would not wish to receive. There has been concern from some patient groups with

neuromuscular disorders that if treatment is withdrawn due to a judgement about the quality of their life, this might not match their own perceived quality of life. It is clear that suitable advance directives stating which treatments they wish to receive may also be helpful. In the UK, a recent High Court judgement has confirmed that doctors need not provide futile care to patients.

Symptom relief

For relief of major symptoms, such as breathlessness, it goes without saying that airflow obstruction and hypoxaemia should be optimally treated according to evidence-based guidelines. Individuals should be carefully evaluated for common co-pathologies, such as left ventricular dysfunction, uncontrolled atrial fibrillation or anaemia, which are readily addressed. A recent meta-analysis [8] has assessed the role of opioids in the palliation of breathlessness in terminal illness. Of the 18 studies analysed, of which nine concerned nebulised opioid, JENNINGS *et al.* [8] found a small effect on reduction in breathlessness using non-nebulised opioids, but no overall beneficial effects on exercise tolerance or improvement in dyspnoea using nebulised delivery. Despite this, there are sporadic case reports on the successful use of nebulised opioid, *e.g.* nebulised fentanyl, for the palliation of dyspnoea in a cystic fibrosis patient with terminal lung disease in whom a reduction in air hunger and Borg score was noted [9]. An individual approach that carefully considers the bioavailability of the opioid and side-effects is clearly crucial. In some situations, side-effects such as sedation and constipation will outweigh benefit, and these considerations will also depend on whether treatment has been given in the last few days/hours of life or as a longer-term strategy.

Oxygen is commonly prescribed to reduce breathlessness, and should be used to correct hypoxaemia. There is little evidence that short-burst oxygen use is of benefit in COPD. STEVENSON and CALVERLEY [10] have shown that oxygen used on recovery from exercise in patients with severe COPD reduced the duration of dynamic hyperinflation, but, interestingly, this physiological gain did not decrease the perception of breathlessness or speed of recovery.

Pharmacological approaches are also valuable in treating depression, panic attacks and insomnia, all of which may exacerbate dyspnoea and/or reduce the quality of life. Serotonin

selective re-uptake inhibitors may be particularly helpful in depression, panic and anxiety [11]. However, cognitive therapy and exploration of patient fears are important too. An overemphasis on drugs should be avoided. Discussion, reassurance and practical support, such as wheelchair, stair lifts and help for the main carer, should not be underestimated. Few hospice admissions are for end-stage lung disease, but hopefully this situation and the availability of respite care will improve [11].

NIV as a palliative tool

While much attention has been directed at the use of NIV to reduce mortality in acute exacerbations of COPD, early work has also shown that NIV reduces dyspnoea in this situation [12], and, therefore, NIV can be reasonably targeted towards symptom relief (figure 2). However, evidence of benefit is less established in steady-state circumstances. Here, careful attention should be focused on the aim of mechanical ventilation. This may be specifically to reduce dyspnoea, ameliorate symptoms related to hypercapnia, such as headaches and poor sleep quality, or potentially to buy time, *e.g.* for an individual to resolve family affairs or to allow time for a distant relative to attend the bedside. In all these situations, the goal of NIV should be decided in advance, so it should be quickly evident whether that aim has been achieved, or if the burden of NIV is outweighing any advantage. There have been few studies in this area, but CUOMO *et al.* [13] have shown that NIV significantly improved oxygenation and Borg dyspnoea score in patients with end-stage solid cancer who develop acute respiratory failure. pH was only improved in hypercapnic patients. NIV can, therefore, be used reasonably to treat this group of patients in whom invasive ventilation is felt to be inappropriate [14].

There is increasing information on the use of NIV in end-stage neuromuscular conditions, such as motor neurone disease/amyotrophic lateral sclerosis. While uncontrolled studies consistently



Figure 2
A patient using NIV during an acute exacerbation of COPD.

suggest NIV can increase survival and improve quality of life [15–17], the extent to which patients with bulbar-onset disease benefit is less clear, particularly as NIV may be more difficult to initiate and tolerate in this group [18]. However, a recent randomised controlled trial has confirmed that NIV improves survival and quality of life in motor neurone disease patients with mild or moderate bulbar disease, and although survival was not prolonged in those with severe bulbar involvement, a range of quality of life measures did improve [19].

Conclusion

All patients with chronic respiratory disease will ultimately enter a terminal phase unless they die of another cause. Care will improve with increasing acknowledgement by physicians and patients that this phase, although inevitable, is not untreatable. In general, patients value discussion of their treatment management and this needs to focus on the life that remains, as well as end-of-life planning. Simple direct questions such as "How do you see your chest problems developing in the future?" and "I'd like to know your views on how you would like to be treated if your condition deteriorated suddenly" should pave the way to a more open, rewarding and mutually beneficial patient–physician relationship.

Educational questions

Are the following statements true or false?

1. Short-burst oxygen therapy reduces dyspnoea in end-stage lung disease.
2. Following an acute exacerbation of COPD requiring NIV, ~20% of patients are admitted to hospital in the following year.
3. An important physician barrier to the discussion of advance directives is a lack of time.
4. NIV is contraindicated as a palliative tool in COPD.

Suggested answers

1. False
2. False
3. True
4. False

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