OCCASIONAL REVIEW

Ethics and decision making in end stage lung disease

A K Simonds

Thorax 2003;**58**:272–277

Most physicians believe they do more good than harm, and these duties of helping and not harming the patient are rooted in the Hippocratic oath, the good Samaritan tradition, and the Order of the Knight Hospitallers founded in the 11th century to care for pilgrims and those wounded in the Crusades.[1] In recent times the simple principles of beneficence and non-maleficence have been augmented and sometimes challenged by a rising awareness of patient/consumer rights, and the public expectation of greater involvement in medical, social and scientific affairs which affect them. In a publicly funded healthcare system in which rationing (explicit or otherwise) is inevitable, the additional concepts of utility and distributive justice can easily come into conflict with the individual's right to autonomy. Possible treatment options for end stage lung disease include transplantation and long term invasive ventilation which are challenging in resource terms. Other interventions such as pulmonary rehabilitation and palliative care are relatively low cost but not uniformly accessible.

Moral or value judgements are made daily in clinical practice, but in respiratory medicine some of the most difficult ethical decisions involve individuals with end stage lung disease. This review is not comprehensive but focuses on the use of mechanical ventilation in severe chronic obstructive pulmonary disease (COPD) and progressive neuromuscular disease, withholding and withdrawing treatments, and end of life decision making. For any intervention, determining the balance between beneficence and maleficence requires an up to date understanding of what that intervention can achieve for the individual and the burdens it will impose. Good communication between physician and patient is essential so that rational decisions can be reached mutually. As there have been advances and recent debate in many of these areas, outcome information is discussed and existing guidelines are highlighted.

Correspondence to:
A K Simonds, Sleep & Ventilation Unit, Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK; a.simonds@rbh.nthames.nhs.uk

OUTCOME OF MECHANICAL VENTILATION IN ACUTE EXACERBATIONS OF COPD

There is a general perception that the outcome of mechanical ventilation in patients with COPD is poor. This expectation has been partly modified in recent years with ICU mortality rates of 11–35% being quoted.[2] In an Australian retrospective study[3] 79.7% of COPD patients admitted to the ICU with an acute exacerbation survived to discharge, with a mortality at 1 year of 48.6%. While it is known that patients with COPD who require prolonged ventilation (>72 hours) or reintubation have a worse prognosis,[2] Breen *et al*[3] found that the median requirement for ventilatory support was 2 days (mean 3.2 days) and only 13% received ventilatory support for more than 1 week—a finding contrary to the perception that weaning problems are common. The mean forced expiratory volume in 1 second (FEV_1) for the group (n=74) was 0.74 ml and, although FEV_1 predicted the need for intubation (presumably as an index of respiratory reserve), neither FEV_1 nor functional performance scores were significant determinants of long term survival. In a larger retrospective series[2] of COPD patients receiving invasive ventilation in the ICU, overall hospital mortality was 28% but this fell to 12% in COPD patients without co-morbidity. Users of long term oxygen therapy (LTOT) did not have a worse outcome than those who were not using oxygen before admission. In a prospective multicentre study Seneff and coworkers[4] found that the presence of non-respiratory organ failure was the most significant predictor of both hospital mortality and 6 month survival (total explanatory power on multivariate analysis of 60% and 54%, respectively). It therefore seems that the presence of comorbidity over and above other prognostic features is the single most useful predictor of survival. Age was also a factor, the hospital mortality rate of 24% rising to 30% in those >65 years old. This, too, may reflect age related development of additional pathology.

Impact of new treatments

An important consideration is that, even in the most recent studies of mechanical ventilation in COPD, non-invasive positive pressure ventilation (NIV) was either unavailable[3] or used in only a small number of patients.[2] Randomised controlled studies[5 6] have now shown that the use of NIV in acute exacerbations of COPD reduces mortality and the need for ICU admission and intubation. Furthermore, early application of NIV facilitates weaning in patients with COPD.[7] Most of the improvement in outcome can probably be attributed to avoiding the complications of intubation such as nosocomial pneumonia. With the increasing availability of NIV, it is likely that the mortality rate for patients with COPD in the ICU may increase as individuals treated with invasive ventilation will be those too sick for NIV (for example, comatose or with multisystem failure) or those in whom NIV has failed (a group in whom the prognosis is poorer anyway).[8] It is important that, at the time of initiation of NIV, a decision is made as to whether progression to

intubation and intermittent positive pressure ventilation (IPPV) is appropriate if NIV is unsuccessful.

Burden of mechanical ventilation

Despite relatively good short term outcomes in COPD patients without comorbidity receiving mechanical ventilation, it should be remembered that the burden of ICU care for the individual is substantial. Over and above medical complications and their consequences, Pochard *et al*[9] found that, in 43 patients receiving IPPV, 88% were depressed, 58% felt unable to communicate, 37% experienced an acute fear of dying, 30% felt intolerable pain, and 21% feared they had been abandoned. Notwithstanding the crucial need to address these issues within the ICU, it is probable that some physical and psychological effects will never be eliminated. In a UK study[10] that reported a 1 year mortality rate of 59% following mechanical ventilation in COPD, 53% of survivors were dependent on carers and were housebound, and their general practitioners felt that 59% of survivors had a greater dependence on carers, worse exercise tolerance, and a poorer quality of life than before ICU admission.

The experience of end stage COPD

Lynn *et al*[11] have looked at the experiences of patients with COPD who died within 1 year of an index hospital admission, focusing particularly on the last 6 months of life. In this group 75% described their quality of life as fair or poor. Around 20% of the final 6 months was spent in hospital. On average each individual had 2–3 comorbidities and 84% had one other comorbidity. Mortality and hospital admissions were closely related to non-respiratory organ failure. Doctors tended to underestimate the survival of patients with the most severe COPD. In the patients themselves survival estimates were inclined to be related to the individual's assessment of their quality of life, but estimates of their prognosis by patients with end stage disease were poorly correlated with actual survival. This creates the potential for conflict between patients and medical teams when determining the appropriateness of interventions, and may impact upon discussions regarding medical futility.

END OF LIFE DECISION MAKING

Ideally, medical decisions should be made by the patient, his family and physician jointly, basing these on the patient's goals, prognosis and the physician's judgement.[12] Most work suggests that communication in this area is inadequate. Wenger *et al*[12] evaluated physician understanding of patient preferences for cardiopulmonary resuscitation (CPR) in over 5000 seriously ill patients in whom around a third had COPD or acute respiratory failure; 64% of patients wanted to receive CPR and 36% wished to forego it. In general, physicians correctly understood 86% of patient preferences *for* CPR, but only 46% of patient preferences *not* to receive CPR. Physicians were more likely to understand a patient preference to forego CPR if they had known the patient for longer, perceived the patient's quality of life to be low, and predicted poor 6 month survival. As a consequence of this mismatch of expectations, 42 patients whose physicians misunderstood their preference to forego CPR underwent a resuscitation attempt (34 of these died before leaving hospital). These results indicate clearly that patients are able to make decisions, but these are not always discussed with the healthcare team. This may be due to fear, anxiety about removing hope on both sides, but also a failure to provide the opportunity for discussion and information exchange. Golin *et al*[13] looked prospectively at communication of resuscitation preferences during a 2 month hospital admission and found that 30% of individuals communicated a preference over this period, and this was most likely to happen if the preference was to forego CPR or to change to foregoing CPR. However, only 50% who had formulated a decision had communicated this to their physician. Strangely, those who expressed a desire to take part in decision making were not more likely to impart this information, and there was no relationship between declining quality of life or ability to carry out activities of daily living and the desire to communicate wishes. Even among those who had completed advance directives before entering hospital, 63% had not communicated these to their physician, implying a continuing degree of conflict between making decisions and the reality of these being acted upon. In view of these misunderstandings, perhaps the best way forward is to create an open channel for the regular discussion of the individual's hopes, fears, and expectations for symptom management and evolving end of life care, rather than focusing primarily on their death.[13]

Looking at the situation from the point of view of the physician, Hinkka *et al*[14] found that physician decisions to forego life prolonging treatment were correlated with experience but, in addition, supervision and postgraduate training in this area were positively associated. With regard to patient related factors which inform physician decision making, the most important were physician estimates of the patient's global quality of life, physical comfort, mobility and depression[15] but, in general, these estimates tended to be worse than the patient's own estimate of his/her quality of life, especially for older patients. Furthermore, patients' own ratings of their quality of life did not correlate well with their preferences for life sustaining interventions.

Protocols and guidelines for withholding and withdrawing life prolonging treatment

There are now clear guidelines on withholding and withdrawing life prolonging treatment, resuscitation, and DNR policies in the US, Europe, and the UK. In the UK, British Medical Association (BMA)[16] and General Medical Council (GMC) guidelines[17] are helpful sources of reference. All emphasise that the primary goal of treatment is to restore or maintain health, but that if treatments fail or adverse effects outweigh benefits, then the justification for providing that treatment is lost. Treatment which does not provide net benefit—that is, is futile—may be ethically and morally withheld or withdrawn, and there is no legal or moral difference between withholding and withdrawing treatment. Voluntary refusal of life prolonging treatment must be respected in adults with decision making capacity. Where individuals have lost the capacity to make decisions, a valid advance directive (written or oral) refusing life prolonging treatment must be respected. At present in England, Wales and Northern Ireland no surrogate has the power to give or withhold consent for an adult who lacks decision making capacity (although the situation may change), but in Scotland a proxy decision maker can be appointed.

Medical futility

Futility is used to denote the absence of benefit or "uselessness" and is derived from the Greek legend in which the daughters of Danaus were condemned to collect water for eternity in leaking buckets as a punishment for murdering their husbands. While futility is generally understood to indicate that an intervention is highly unlikely to produce meaningful survival, some ethicists[18] have argued that futility is used as a trump card to overrule the autonomous wishes of patients, and that harm results from failing to respect the patient's autonomy. However, this harm is outweighed by embarking on a treatment which produces no benefit, and practice guidelines free the physician from any obligation to provide a futile medical treatment. Clearly the physician must be as sure as possible in his own mind that an intervention is overwhelmingly unlikely to produce net benefit.

What happens in practice?

There appear to be widespread variations in clinical practice despite the ubiquity of guidelines.

In a European study[19] involving 16 countries, 73% of ICUs admitted patients with no hope of survival (although 33% of respondents felt such patients should be admitted), and 80% felt that DNR orders should be used, but only 58% did so (ranging from 8% in Italy to 91% in the Netherlands). 93% would withhold treatment, but withdrawal was less common. Just under 50% of units included patient, staff, and family in decision making.

In the UK Ravenscroft and Bell[20] have reported substantial variations in practice with regard to life sustaining care, although their survey of Yorkshire ICUs was carried out before publication of the BMA guidelines. The ICUs reported inconsistency in admissions policy and DNR policy and eight of the 13 units perceived that decisions on the withdrawal of treatment were inconsistent. The reasons cited for inconsistency included family pressure, decisions based on an arbitrary quality of life judgements, and active treatment continued despite a nursing perception of futility. Inconsistencies in the withdrawal of treatment were reported to be particularly notable in cases with similar pathology, such as COPD.

What can be done?

Process of decision making

Guidelines should be consulted and applied where available. Many hospital trusts provide local versions. The inherent uncertainty in medical practice should be acknowledged. Health professionals have an ethical obligation to keep up to date and base decisions on the most accurate information available. A second opinion should be sought where expertise and experience is lacking, or there is disagreement on management. Where there is reasonable doubt about the potential for benefit, a treatment can be provided for a pre-agreed period, subject to review at the end of that period. Before a decision to withhold or withdraw treatment is made, adequate time, resources and facilities should be available for a thorough assessment to obtain information from the patient on preferences, goals and self-reported quality of life, where possible. A consensus between staff, patient, and family should be the aim. If the clinician's view is challenged or conflict arises, a second opinion, independent ethics consultation or, as a final resort, review by the courts is advisable. Ethics consultations in the ICU have recently been shown to reduce the duration of stay and the time on aggressive life sustaining treatments, without changing mortality.[21]

Withdrawal of treatment is more accurately described as "redirection of care"[22] as immediate steps should be taken to institute palliative care measures which may include sedation and analgesia. The principle of "double effect" permits the relief of suffering by such medication despite the fact that life may be shortened as a result. Helping to achieve a "good death" is an entirely appropriate clinical aim and will include attention to clear decision making, symptom management, affirmation of the individual, strengthening relationships with loved ones, relieving burden, and avoiding prolongation of the dying process.[23–25]

Facilitating decision making for patients

End of life decision making can only be improved if opportunities for discussion exist. The notion that these issues should not be explored unless raised by the patient is unsubstantiated. Focus group discussions have shown that patients with COPD in the US wish to be educated on all aspects of their disease, and feel that information on short and long term prognosis and advance directives is often deficient.[26] A similar lack of information has been reported by COPD patients in the UK.[27] Discussion during pulmonary rehabilitation sessions is a useful way of raising these issues, and certainly such debate could be incorporated fairly easily into a standard pulmonary rehabilitation programme. Individual exploration and follow up of issues is then required as the extent of information required by each individual and the values attached to it will clearly vary. Unfortunately, not all COPD and non-COPD patients who would benefit from pulmonary rehabilitation have access to such programmes. Furthermore, palliative care services which help individuals to address end of life issues are poorly developed and fragmented for patients with chronic disorders such as COPD and congestive heart failure compared with the fairly comprehensive provision for those with malignant disease.

Most would accept that patient preferences on specific issues such as intubation and mechanical ventilation or CPR can be influenced by the way in which information on the intervention and its consequences is presented.[28] In an attempt to improve decision making on intubation and mechanical ventilation in patients with COPD, Dales *et al*[29] have developed a standardised scenario-based aid consisting of an audio cassette and book describing intubation, mechanical ventilation (MV) and its possible outcomes, and applied this aid in 20 patients with COPD (10 men) with FEV_1 <45% predicted. Five had received mechanical ventilation previously. Before using the decision aid 50% chose to forego MV, 35% wished to receive it, and three were undecided. After using the aid, 60% wished to forego MV, 40% chose to receive it, and none were undecided. Interestingly, all 10 women declined MV and, although scores on the SF 36 quality of life scale were slightly higher in those who wished to receive MV, this trend did not reach significance. There was congruence between physician and patient decisions in 65% of cases, but agreement between surrogate understanding of the patient's wishes and the patient's own decision was extremely poor. The gender issue may be important to examine further, as in other areas of health care women have been shown to be less likely to take up active options.[30] The authors[29] suggest that women and men may weigh criteria differently in the decision making process—for example, women may have a stronger desire not to burden their family or may be more risk averse. This has implications for the most helpful way to frame information.

As outlined in the BMA and GMC guidelines, competent adults have the right to decide how much weight to attach to the benefits, burdens, risks, and overall acceptability of treatment. Often the outcome of interventions is discussed with patients, but there is little emphasis on the burdens of treatment. Fried *et al*[31] have assessed how treatment preference is affected by the burden of treatment, as well as the outcome, in groups of patients with either COPD, congestive heart failure, or cancer. The burden of treatment included aspects such as length of hospital stay, extent of testing, and invasiveness of interventions. The results showed that the burden of treatment had a significant impact on decision making—for example, 98.7% of participants would accept a low burden treatment if it restored current levels of health, but 11.2% would refuse high burden treatment that produced a similar outcome. For the three age matched diagnostic groups, patients with COPD had the worst perceived health and functional levels but there was no difference in treatment preferences. Patients were also able to assign a value to outcomes other than death, with over 70% stating that they would reject low burden treatment that resulted in severe functional or cognitive impairment. As might be expected, the proportion electing to receive treatment declined as the likelihood of an adverse outcome increased. These data were presented in "pie chart" form and, contrary to suggestions that such concepts would be difficult to understand, the results show that the participants were able to incorporate probabilistic thinking into their decision making.

Advance directives

There is no legislation in England and Wales governing advance directives or living wills, but they are recognised by

case law and, as indicated above, doctors are expected to comply with them. This provides many individuals with comfort and reassurance that the care they receive will be in accordance with their wishes. Knowledge about advance directives is not widespread but, in a survey of elderly people living in London, a large majority (74%) expressed an interest in writing a living will.[32] A poll in 2000 showed that over a quarter of hospital trusts in the UK had developed or intended to develop a policy to encourage advance directives, and this number is likely to grow.[33]

RESPIRATORY COMPLICATIONS OF PROGRESSIVE NEUROMUSCULAR DISEASE

A further area where ethical issues frequently arise for respiratory physicians is in the management of patients with neuromuscular disease or neurological disorders which impair respiratory function. This is especially the case in Duchenne muscular dystrophy (DMD), motor neurone disease/amyotrophic lateral sclerosis (MND/ALS), and children with severe spinal muscular atrophy (SMA). In all these conditions respiratory complications produce burdensome symptoms and are the most common cause of death. Without ventilatory support the average age of death in patients with DMD is 18–20 years,[34] and almost all children with type 1 SMA die by the age of 2 years.[35] The prognosis is less than 1 year in patients with DMD who have become hypercapnic.[36] While long term mechanical ventilation has been applied sporadically for these patient groups for decades, it is only in recent years that the overall benefit from home ventilatory support has been systematically evaluated in larger numbers of patients in terms of survival, palliation of symptoms, and quality of life. Also, the advent of new modes of non-invasive ventilation (NIV) has widened (and probably simplified) the options available. At present restoration of health is not possible in these incurable conditions, but the general principle of beneficence dictates that the physician should do all he can to palliate symptoms and maintain or even prolong a quality of life that is acceptable to the individual. For example, NIV in patients with DMD can result in a 5 year survival of over 70% with good self-reported quality of life.[37] Several studies of NIV in patients with MND/ALS with respiratory insufficiency have shown prolonged survival[38–40] and an improved quality of life in the face of continued functional decline,[41 42] although patient selection is important. Those with early respiratory muscle involvement, sleep related symptoms, orthopnoea, and fewer bulbar problems are most likely to benefit.[43] Nearly all neuromuscular patients treated with NIV opt to continue it, even in the terminal phase of the disease. At the same time, interventions which cause distress, limit privacy, and impose burdens do not conform with the principle of non-maleficence. Assisted ventilation can clearly be both beneficent and maleficent and, indeed, may be clinically (if not physiologically) futile in a quadriplegic, locked-in individual with MND/ALS. However, patients with DMD and MND/ALS usually retain normal capacity for decision making (although assistance with communication may be required), and their autonomy should be respected. Competent patients have the right to refuse life sustaining treatment or request for it to be withdrawn, and their wishes should be honoured. In a case series of patients with MND/ALS patients receiving long term ventilation, 76% completed advance directives and 96% approved of them.[44] Most patients wished to set limits to ventilation in certain circumstances—for example, becoming locked in—and felt that advance care planning facilitated communication of their preferences to family and physicians.

Fully informed decisions are not possible, though, if information on treatment possibilities is withheld. Of concern is the fact that the option of mechanical ventilation may not be discussed with some patients with progressive neuromuscular conditions in the belief that this intervention would be likely to prolong suffering. Bach[45] found that, of 273 directors of Muscular Dystrophy Association clinics in the USA, 41% discouraged ventilator use in muscular dystrophy patients and 41% of these clinicians justified this decision citing "poor quality of life". Only two physicians who discouraged ventilator use were familiar with non-invasive modes of ventilation. Physician beliefs and practices regarding long term ventilation in patients with DMD have recently been examined in a Canadian study[46] which showed that 25% of physicians who care for patients with DMD did not discuss assisted ventilation with all patients. Here, too, 52% of physicians cited a resulting poor quality of life as the reason for not discussing or advising against ventilatory support. However, in the interests of autonomy it is crucial that unilateral decisions based on physician judgements of quality of life are avoided, not least because healthcare professionals consistently underestimate the quality of life of muscular dystrophy patients/ventilator users.[47]

Use of NIV raises the new concern of whether increasing ventilatory dependence and bulbar involvement should lead patients to progress routinely to tracheostomy ventilation. In some countries such as France this would be seen as a natural progression and part of a continuing care plan. In the UK the relative lack of home care services makes the discharge of a tracheostomised ventilator dependent patient a complex and fairly lengthy procedure. It should also be taken into account that recent outcome data[48] show that a non-invasive approach may result in less pulmonary morbidity than tracheostomy ventilation in patients with DMD, and some patients may wish to set a ceiling to their level of care at NIV. The argument to withhold information on invasive ventilation is difficult to sustain, however. In each individual there will be a balance between autonomy, beneficence (palliation of symptoms and extension of life), and maleficence (risk of surgery, prolonged hospitalisation, loss of normal speech, reduced privacy due to increased care package) which needs to be set against the further principle of distributative justice.

If anything, considerations become more stark in the management of infants born with type 1 SMA. Nearly all affected babies develop recurrent respiratory tract infections and respiratory failure within the first few months of life. Infants with type 1 SMA are differentiated functionally from those with type 2 disease by an inability to sit. As in MND/ALS,[49] there are clear cultural differences in providing mechanical ventilation to these profoundly weak children. In Japan[50 51] and France[52] children with type 1 SMA are likely to be treated with tracheostomy ventilation, whereas in the UK this is almost never provided. Indeed, in 1997 the UK High Court approved of doctors' plans to withdraw artificial ventilation and not to resuscitate a 16 month old infant with type 1 SMA which was against the wishes of the parents.[53] Since that case, several publications have reported the outcome of NIV in type 1 SMA. One study showed that prolonged survival was possible but did not give information on quality of life,[54] and another showed no meaningful effect on life expectancy, although it was possible for the children to die at home.[55] Despite these new developments, it remains the case that, in the most severely affected SMA infants, control of ventilation using non-invasive modes is impossible. It is also probable that the study[55] showing significant improvement in prognosis predominantly included less severely affected type 1 children, as the average age of development of respiratory complications was very late at 14 months. This illustrates that blanket decisions based on a diagnostic label are not helpful. There is a fairly wide variation of functional ability within the type 1 spectrum such that Dubowitz[56] has advocated a classification of type 1.0–1.9, 2.0–2.9, etc. A child with type 1.9 SMA therefore has more in common functionally with a child with type 2.1 SMA (in whom ventilatory support is less controversial) than a child with type 1.1 SMA. Individual assessment is therefore critical before a decision regarding ventilatory support is made.

AUTONOMY AND THE CASES OF MS B AND DIANE PRETTY

The obvious similarities between these well publicised cases are that both concerned individuals who were paralysed from the neck down and wished to end their life, and court decisions were required to resolve conflict between the autonomy of the individual and the responsibilities of their physicians. However, the essential difference between the cases is that Ms B was seeking withdrawal of assisted ventilation, whereas Mrs Pretty who had MND/ALS was requesting assistance to end her life. The court held that Ms B was competent and entitled to end her life by refusing to continue life support. Several weeks later the European Court of Human Rights rejected an application by Mrs Pretty who claimed that a UK High Court decision which refused to allow her husband to assist her suicide contravened articles 3, 8 and 14 of the Convention for the Protection of Human Rights and Fundamental Freedoms Act. From a purely legal point of view, the outcome of these cases was not surprising—Ms B was judged competent and therefore her autonomy was respected and ventilation was withdrawn. However, the European Court of Human Rights found that Mrs Pretty was not being subject to inhuman or degrading treatment, nor were her rights to autonomy being infringed, and she was not being discriminated against on the grounds of being physically disabled. Her problem was that, unlike Ms B, the disease rather than medical intervention created the predicament she found herself in. The judges concluded that Mrs Pretty did not have the right to challenge established English law. Despite these findings, Singer[57] has argued that philosophically there is little difference between the cases: "we have arrived at the absurd situation where a paralysed woman can choose to die when she wants if her condition means she needs some form of medical treatment to survive; whereas another paralysed woman cannot choose to die when or in the manner she wants because there is no treatment keeping her alive in such a way that, if it were withdrawn, she would have a humane and dignified death. What we have done is build legal doctrines based on two separate rules of law, and thereby we have reached a situation that makes no ethical sense at all. We need to move beyond a rule-based ethic, and consider the consequences of the situations in which we are faced."

However, the currently prevailing view is that the benefit to the individual of having their wish for euthanasia or assisted suicide respected can only be achieved at too great a cost to society. Active or intentional termination of another person's life remains morally and legally different from withdrawal of treatment and contravenes the law. Public airing of these difficult issues is likely to be helpful long term, and this debate will continue to be informed by developments in the Netherlands and Oregon where assisted suicide is sanctioned and regulated.

REFERENCES

1 **Randall F**, Downie RS. Ethics and aims in palliative care. In: Randall F, Downie RS, eds. *Palliative care ethics. A good companion*. Oxford: Oxford University Press, 1996: 1–24.
2 **Nevins ML**, Epstein SK. Predictors of outcome for patients with COPD requiring invasive mechanical ventilation. *Chest* 2001;**119**:1840–9.
3 **Breen D**, Churches T, Hawker F, *et al*. Acute respiratory failure secondary to chronic obstructive pulmonary disease treated in an internsive care unit: a long term follow up study. *Thorax* 2002;**57**:29–33.
4 **Seneff MG**, Wagner DP, Wagner RP, *et al*. Hospital and 1-year survival of patients admitted to intensive care units with acute exacerbation of chronic obstructive pulmonary disease. *JAMA* 1995;**274**:1852–7.
5 **Brochard L**, Mancebo J, Wysocki M, *et al*. Noninvasive ventilation for acute exacerbations of chronic pulmonary disease. *N Engl J Med* 1995;**333**:817–22.
6 **Plant PK**, Owen JL, Elliott MW. Early use of noninvasive ventilation for acute exacerbations of chronic obstructive pulmonary disease on general respiratory wards: a multicentre randomised controlled trial. *Lancet* 2000;**355**:1931–5.
7 **Nava S**, Ambrosino N, Clini E, *et al*. Non-invasive mechanical ventilation in the weaning of patients with respiratory failure due to chronic obstructive pulmonary disease. A randomized controlled trial. *Ann Intern Med* 1998;**128**:721–8.
8 **Moretti M**, Cilione C, Tampieri A, *et al*. Incidence and causes of non-invasive mechanical ventilation failure after initial success. *Thorax* 2000;**55**:819–25.
9 **Pochard F**, Lanore JJ, Bellivire F, *et al*. Subjective psychological status of severely ill paitents discharged from mechanical ventilation. *Clin Intensive Care* 1995;**6**:57–61.
10 **Hill AT**, Hopkinson RB, Stableforth DE. Ventilation in a Birmingham intensive care unit 1993–1995: outcome for patients with chronic obstructive pulmonary disease. *Respir Med* 1999;**92**:156–61.
11 **Lynn J**, Ely EW, Zhong Z, *et al*. Living and dying with chronic obstructive pulmonary disease. *J Am Geriatr Soc* 2000;**48**: S91–100
12 **Wenger NS**, Phillips RS, Teno JM, *et al*. Physician understanding of patient resuscitation prefernces: insights and clinical implications. *J Am Geriatr Soc* 2000;**48**: S44–51
13 **Golin CE**, Wenger NS, Liu H, *et al*. A prospective study of patient-physician communication about resuscitation. *J Am Geriatr Soc* 2000;**48**: S52–60
14 **Hinkka H**, Kosunen E, Metsanoja R, *et al*. Factors affecting physicians' decisions to forgo life-sustaining treatments in terminal care. *J Med Ethics* 2002;**28**:109–14.
15 **Uhlmann RF**, Pearlman RA. Perceived quality of life and preferences for life-sustaining treatment in older adults. *Arch Intern Med* 1991;**151**:495–7.
16 **British Medical Association**. *Withholding and withdrawing life prolonging medical treatment: guidance for decision making*. London: BMJ Publishing Group, 1999.
17 **General Medical Council**. *Withholding and withdrawing life-prolonging medical treatment: good practice in decision-making*. London: General Medical Council, 2002.
18 **Ardagh M**. Futility has no utility in resuscitation medicine. *J Med Ethics* 2000;**26**:396–9.
19 **Vincent J-L**. Forgoing life support in western European intensive care units: the results of an ethical questionnaire. *Crit Care Med* 1999;**27**:1626–33.
20 **Ravenscroft AJ**, Bell MDD. 'End-of-life' decision making within intensive care - objective, consistent, defensible? *J Med Ethics* 2000;**26**:435–40.
21 **Schneiderman LJ**, Gilmer T, Teetzel HD. Impact of ethics consultations in the intensive care setting: a randomised, controlled trial. *Crit Care Med* 2000;**28**:3920–4.
22 **Branthwaite MA**. Ethical and medico-legal aspects of assisted ventilation. In: Simonds AK, ed. *Non-invasive respiratory support: a practical handbook*. 2nd ed. London: Arnold, 2001: 282–91.
23 **Singer PA**, Martin DK, Kelner M. Quality end-of-life care: patients' perspectives. *JAMA* 1999;**281**:163–8.
24 **Steinhauser KE**, Clipp EC, McNeilly M, *et al*. In search of a good death: observations of patients, familes and providers. *Ann Intern Med* 2000;**132**:825–32.
25 **Sensky T**. Withdrawal of life sustaining treatment. *BMJ* 2002;**325**:175–6.
26 **Curtis JR**, Wenrich MD, Carline JD, *et al*. Patients' perspectives on physician skill in end-of-life care. Differences between patients with COPD, cancer and AIDS. *Chest* 2002;**122**:356–62.
27 **Gore JM**, Brophy CJ, Greenstone MA. How well do we care for patients with endstage chronic obstructive pulmonary disease (COPD)? A comparison of palliative care and quality of life in COPD and lung cancer. *Thorax* 2000;**55**:1000–6.
28 **Percy ME**, Llewellyn-Thomas H. Assessing preferences about the DNR order: does it depend on how you ask? *Med Decis Making* 1995;**15**:209–16.
29 **Dales RE**, O'Connor A, Hebert P, *et al*. Intubation and mechanical ventilation for COPD. Development of an instrument to elicit patient preferences. *Chest* 1999;**116**:792–800.
30 **Dempsey SJ**, Dracup K, Moser DK. Womens' decisions to seek care for symptoms of acute myocardial infarction. *Heart Lung* 1995;**24**:444–56.
31 **Fried TR**, Bradley E, Towle VR, *et al*. Understanding the treatment preferences of seriously ill patients. *N Engl J Med* 2002;**346**:1061–6.
32 **Schiff R**, Rajkumar C, Bulpitt C. Views of elderly people on living wills: interview study. *BMJ* 2000;**320**:1640–1.
33 **Diggory P**, Judd M. Advance directives: questionnaire survey of NHS trusts. *BMJ* 2000;**320**:24–5.
34 **Rideau Y**, Gatin G, Bach J, *et al*. Prolongation of life in Duchenne's muscular dystrophy. *Acta Neurol* 1983;**5**:118–24.
35 **Thomas NH**, Dubowitz V. The natural history of type I (severe) spinal muscular atrophy. *Neuromusc Disord* 1994;**4**:497–502.
36 **Vianello A**, Bevilacqua M, Salvador V, *et al*. Long-term nasal intermittent positive pressure ventilation in advanced Duchenne's muscular dystrophy. *Chest* 1994;**105**:445–8.
37 **Simonds AK**, Muntoni F, Heather S, *et al*. Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy. *Thorax* 1998;**53**:949–52.
38 **Pinto AC**, Evangelista T, Carvalho M, *et al*. Respiratory assistance with a non-invasive ventilator (BiPAP) in MND/ALS patients: survival rates in a controlled trial. *J Neurol Sci* 1995;**129**(Suppl):19–26.
39 **Kleopa KA**, Sherman M, Bettie N, *et al*. BiPaP improves survival and rate of pulmonary function decline in patients with ALS. *J Neurol Sci* 1999;**164**:82–8.
40 **Bach JR**. Amyotrophic lateral sclerosis. Prolongation of life by noninvasive respiratory aids. *Chest* 2002;**122**:92–8.
41 **Lyall RA**, Donaldson N, Fleming T, *et al*. A prospective study of quality of life in ALS patients treated with non-invasive ventilation. *Neurology* 2001;**57**:153–6.

42 **Polkey MI**, Lyall RA, Davidson AC, *et al*. Ethical and clinical issues in the use of home non-invasive ventilation for the palliation of breathlessness in motor neurone disease. *Thorax* 1999;**54**:367–71.
43 **Aboussouan LS**, Khan SU, Meeker DP, *et al*. Effect of noninvasive positive-pressure ventilation on survival in amyotrophic lateral sclerosis. *Ann Intern Med* 1997;**127**:450–3.
44 **Moss AH**, Oppenheimer EA, Casey P, *et al*. Patients with amyotrophic lateral sclerosis receiving long-term mechanical ventilation. Advance care planning and outcomes. *Chest* 1996;**110**:249–55.
45 **Bach JR**, Barnett V. Ethical considerations in the management of individuals with severe neuromuscular disorders. *Am J Phys Med Rehabil* 1994;**73**:134–40.
46 **Gibson B**. Long-term ventilation for patients with Duchenne muscular dystrophy. Physicians' beliefs and practices. *Chest* 2001;**119**:940–6.
47 **Bach JR**, Campagnolo DI, Hoeman S. Life satisfaction of individuals with Duchenne muscular dystrophy using long-term mechanical ventilatory support. *Am J Phys Med Rehabil* 1991;**70**:129–35.
48 **Bach JR**, Ishikawa Y, Kim H. Prevention of pulmonary morbidity for patients with Duchenne muscular dystrophy. *Chest* 1998;**112**:1024–8.
49 **Borasio GD**, Gelinas DF, Yanagisawa N. Mechanical ventilation in amyotrophic lateral sclerosis: a cross cultural perspective. *J Neurol* 1998;**245**: S7–12.
50 **Sakakihara Y**, Kubota M, Kim S, *et al*. Long-term ventilator support in patients with Werdnig-Hoffman disease. *Pediatr Int* 2000;**42**: 359–63.
51 **Sakakihara Y**. Ethical attitudes of Japanese physicians regarding life-sustaining treatment for children with severe neurologocal disabilities. *Brain Devel* 2000;**22**:113–7.
52 **Barois A**, Estournet-Mathiaud. Ventilatory support at home in children with spinal muscular atrophies (SMA). *Eur Respir Rev* 1992;**10**:319–22.
53 **Dyer C**. British court allows terminally ill baby to die. *BMJ* 1997;**315**:1397–1402.
54 **Birnkrant DJ**, Pope JF, Martin JE, *et al*. Treatment of type I spinal muscular atrophy with non-invasive ventilation and gastrostomy feeding. *Pediatr Neurol* 1998;**18**:407–10.
55 **Bach JR**, Niranjan V, Weaver B. Spinal muscular atrophy type I. A noninvasive respiratory management approach. *Chest* 2000;**117**:1100–5.
56 **Dubowitz V**. Disorders of the lower motor neurone: the spinal muscular atrophies. In: Dubowitz V, ed. *Muscle disorders in childhood*. 2nd ed. London: W B Saunders, 1995: 325–67.
57 **Singer P**. Ms B and Diane Pretty: a commentary. *J Med Ethics* 2002;**28**:234–5.