

# Issues for palliative medicine doctors surrounding the withdrawal of non-invasive ventilation at the request of a patient with motor neurone disease: a scoping study

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## ABSTRACT

**Background** Non-invasive ventilation (NIV) is beneficial for respiratory failure in motor neurone disease (MND) but some patients may wish to stop the intervention. Guidance from the National Institute for Health and Care Excellence recommends that research is needed on NIV withdrawal. There is little in the literature focusing on the issues doctors face when withdrawing NIV in this group.

**Aim** To identify issues and challenges that palliative medicine doctors encounter in relation to the withdrawal of NIV in MND patients.

**Method** An electronic questionnaire was sent to members of the Association of Palliative Medicine of Great Britain and Ireland.

Participants rated how practically, emotionally and ethically challenging they found the process of NIV withdrawal.

**Results** 76 doctors responding had been directly involved in withdrawal of NIV at the request of a patient with MND. A high percentage rated the practical, ethical and emotional challenges as 7 or more on a 0–10 scale. Thematic analysis of the free text revealed some common difficulties. Lack of guidance on practical aspects of withdrawal, poor advance care planning and the need to support all involved to prevent conflict were recurrent themes. Statements relating to the emotional burden were diverse but suggest many palliative care doctors feel significant personal impact.

**Conclusions** The withdrawal of NIV in patients with MND appears to pose considerable challenges to palliative medicine doctors; emotionally, practically and to a lesser extent ethically. Development of guidelines and a clear ethical statement of conduct may help but emotional issues appear more complex.

## BACKGROUND

Motor neurone disease (MND) is a progressive neurological condition which primarily affects motor neurones leading to skeletal muscle weakness. Respiratory muscle weakness leads to fatigue, breathlessness, hypoxia, hypercapnia, morning headaches, poor sleep, anorexia, anxiety and the risk of infection due to poor cough clearance of secretions. Most deaths in MND are due to respiratory failure. NIV can improve both symptoms and survival,<sup>1,2</sup> and recent UK guidance from the National Institute for Health and Care Excellence (NICE) supports its use.<sup>3</sup>

Some, but by no means all, patients with respiratory failure due to MND choose to use NIV. Little is known about patient decision making but it would seem that many patients make this choice to improve their quality of life. Other work has suggested that the disease itself takes away any sense of autonomy in decision making.<sup>4</sup> Undoubtedly, NIV has many benefits for patients.<sup>5,6</sup> Neurological deterioration is however relentless for patients despite NIV and patients may eventually reach a point at which they cannot move or communicate. Many also develop dementia and cognitive change, primarily frontal lobe dysfunction, which affects their executive functioning and ability to make decisions. The ability to control discontinuation of ventilation may be a crucial factor for the patient making the decision about starting the treatment.<sup>7</sup>

One study found that half of the users of respiratory support discontinued it well before dying from their MND.<sup>8</sup>

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Additional information should shortly be available from two longitudinal cohort studies of patients using NIV which have recently been undertaken in the UK (McDermott and Young, personal communication 2012). There is little other literature that helps us understand what proportion of patients become dependent on NIV (ie, get severely breathless without it within a very short space of time) and utilise it almost 24/7, but retrospective studies suggests this may be so for around 50% of patients who use NIV. NIV can very effectively support ventilation, often for many months, but the majority of patients using NIV 24/7 in the UK appear to die while still using it. In the UK, very few people are on tracheostomy delivered ventilation.

Some patients who are dependent on NIV will wish it to be stopped because they can no longer tolerate it or because of deterioration in other factors in their quality of life. Some will have made a written statement with respect to withdrawal in advance of their losing the ability to communicate (or altered capacity).<sup>9</sup> If properly written and witnessed, an advance decision to refuse treatment (ADRT) is legally binding and clearly states the patient's wishes.<sup>10</sup> Interviews with patients on assisted ventilation in the USA (mostly receiving this via tracheostomy) found that while most patients wanted to issue a direction in advance about circumstances in which they would wish to stop assisted ventilation, few had had the opportunity to do so.<sup>11</sup> Whitehead *et al*<sup>12</sup> explored end of life decision making with patients and with bereaved carers in Preston, UK, and identified a need for more information and shared decision making, but there is little other literature exploring this area in the UK.

The recent UK NICE guidance identifies the lack of evidence on providing information to family and patients using NIV in relation to end of life care and was unable to draw on any evidence pertaining to the timing and content of end of life care discussions with patients and family.<sup>3</sup> The guidance does suggest that discussion of withdrawal should take place before or as NIV is started, and discussions regarding end of life care should be instigated if the patient or family requests this, at this time or as the patient deteriorates.

#### Withdrawal of NIV

The NICE guidance also states that more research is needed on the withdrawal of NIV, identifying the lack of any clarity in the most effective and acceptable method of withdrawal and how this process should be facilitated and managed. Specifically, NICE suggests interviewing the professionals involved in such events as a focus of research (see NICE Clinical Guideline 105<sup>3</sup>, p96).

For those patients who use NIV most of the time, removal of NIV will usually be followed by death within hours, although anecdotally this has proved

surprisingly variable. Removal of NIV usually leads to a rapid onset of distressing symptoms and medication is needed to manage the physical distress that may result from NIV withdrawal. There is a very small literature discussing the clinical and practical aspects of withdrawal of NIV in MND,<sup>3 13–18</sup> and some on the withdrawal of ventilation for other conscious patients.<sup>19–22</sup> Other than a recent BBC radio programme<sup>23</sup> and two case studies,<sup>21 24</sup> there is no work which has looked at what can be learned from the experiences of professionals or close family carers.

Withdrawing NIV support from an MND patient whose life depends on it is a rare event. There are no statistics available but our experiences suggests at most currently 1–2 people/million population per year in the UK or around 0.5–1% of patients with MND. This could increase with the increased awareness of the use of NIV following the NICE guidance and if trials of diaphragmatic pacing show benefit. It is clearly important to learn as much as possible from those that have experience of managing such patients in order to inform practice for the future, especially as large scale studies on this aspect of care are unlikely to be feasible.

#### Impact of NIV withdrawal on health professionals

Experience tells us that all concerned (patient, family and professional caregivers) find stopping something that has been keeping a person alive a difficult situation. However, there is minimal research about the experience and concerns of any party.

There has been an exploration of the ethical context of withdrawal of NIV,<sup>13 14 21 25</sup> and this emphasises the stressful nature of this area of clinical care and how differences in opinion and belief, despite a uniform legal context, can influence a doctor's practice and level of distress. The literature has not explored the actual experiences of doctors (or indeed of patients, their families or other health professionals).

Withdrawal of life sustaining treatment from insentient patients who are dying from organ failure or brain injury (not MND) in intensive care units indicates emotional impact on professional team members,<sup>26</sup> and physicians consider mechanical ventilation the most difficult treatment to withdraw.<sup>27</sup> In the context of MND where patients may remain able to hear, see, think and feel normally, but may not be able to communicate, the impact on their professionals could be expected to be even greater.

Gannon<sup>24</sup> noted a high level of distress for staff and a change in their professional behaviour in a hospice setting where they considered accepting a patient with MND for NIV withdrawal. He postulates that withdrawal of a ventilator generates more concern than withdrawing fluids, for example, since it requires a specific act that will result in death and it is uncommon.

Clinch and Le<sup>21</sup> have recently described withdrawal of mechanical ventilation at home in a conscious patient with irreversible respiratory failure. A feature of this case study was 'the attitudinal barrier encountered from the local domiciliary palliative care service, who were not prepared to be involved directly with the withdrawal of ventilation itself, although they were fully supportive of care of the patient and his family once the procedure had been completed'.

A recent BBC radio 4 *Inside the ethics committee* broadcast further illustrated the issues that can arise through the retelling of the stories of two patients.<sup>23</sup> Issues were raised as to the role of the doctor, the points of uncertainty and the conflicts that may arise in a team when individuals have differing ethical understanding and values.

The Leicestershire guidelines<sup>28</sup> acknowledge that, in view of the complex emotional impact of withdrawing NIV, 'debriefing and support should be available to the professionals involved'. Similarly, report from San Diego comments that a 'debrief for the interdisciplinary group has been found to be essential'.<sup>22</sup>

There is an indication then that not only does the potential or actual withdrawal of NIV have a significant and extraordinary impact on healthcare professionals, it may also have a direct effect on their practice. To achieve best outcomes for patients and their families, the impact on professionals and the support they require to avoid adverse effects on their practice needs to be explored much further.

### Aims

The aim of this study was to explore, from the perspective of doctors in palliative medicine, the issues related to the withdrawal of NIV at the request of a patient with MND and to identify areas for more in-depth exploration.

### METHODS

A mixed categorical and free text questionnaire was developed using the literature and the experience of three consultants in palliative medicine (CF, DO and one other) as a basis for the content. The questionnaire was piloted with registrars in palliative medicine at one hospice, seeking comment and undergoing revision in order to inform reliability and validity.

The opinion of the South East Research Ethics Committee was sought and the chair advised that NRES ethics approval was not required as the study constituted a service evaluation.

The questionnaire was formatted in Survey Monkey and was sent via email to all doctors who were members of the Association of Palliative Medicine of Great Britain and Ireland (APM) (993 doctors). A covering letter outlining the purpose of the study was sent and clearly stated this was a scoping exercise from which further research would be informed. No

reminder was sent. Respondents had 3 months in which to complete the survey.

Participants were asked to rate how practically, emotionally and ethically challenging they found the process of NIV withdrawal on an 11 point Likert scale (0=not at all challenging; 10=very challenging). They were invited to expand on these three areas using free text.

All responses were anonymised. Free text was thematically analysed using constant comparison based on grounded theory.<sup>29</sup> Text was read by CRH and CF and coded independently. Codes were then related to the three major themes (practical, ethical and emotional challenges). Codes were subsequently grouped and collapsed as subthemes emerged (CRH). Subthemes and grouping or allocation of text was discussed between CRH and CF. Categorical data were analysed descriptively.

### RESULTS

A total of 134 people responded, four of whom were not doctors (one physiotherapist, one MND specialist nurse, one clinical nurse specialist in palliative care and one senior nurse). Results are reported here for the 130 questionnaires from doctors involved with MND patients. Eight (6.2%) of the doctors had not cared for a MND patient who used NIV and a further 46 (35.4%) had not been involved in the actual withdrawal of NIV at the request of a patient.

Seventy-six (58.5%) respondents had been directly involved in the withdrawal of NIV, 60% had undertaken this in the past 12 months but 5% had only been involved in withdrawal more than 5 years before. The majority of respondents (59%) had been involved with more than one patient but only 9% had undertaken this in five or more patients in the past 5 years.

The magnitudes of the challenges perceived and experienced by doctors in the practical, ethical and emotional dimensions are shown in table 1. A greater percentage of doctors that had not been involved in

**Table 1** Level of practical, emotional and ethical challenges identified by respondents who had and had not been directly involved in non-invasive ventilation withdrawal at the request of a patient

	Score 0–3	Score 4–6	Score 7+
Those doctors with direct experience of withdrawal of NIV (n (%))			
Practical challenge	17 (22)	27 (36)	32 (42)
Ethical challenge	26 (34)	25 (33)	25 (33)
Emotional challenge	13 (17)	26 (34)	37 (49)
Those doctors involved with MND patients with NIV but no direct experience of withdrawal of NIV (n (%))			
Practical challenge	5 (14)	10 (29)	20 (57)
Ethical challenge	10 (29)	10 (29)	15 (43)
Emotional challenge	8 (23)	7 (20)	20 (57)

MND, motor neurone disease; NIV, non-invasive ventilation.

withdrawal of NIV identified that all of these dimensions presented a level of challenge of 7 or more to them.

### Practical challenges

Only one respondent (who had never been involved in withdrawal of NIV) felt that practical aspects were not at all challenging, but 42% of doctors who had undertaken withdrawal and over half of other doctors scored this as 7 or more on the 0–10 scale. In the absence of guidelines, concerns relating to whether or not to wean ventilation, how to manage distressing symptoms, the use of sedative drugs (what and how) together with who should remove the mask were all issues posed by respondents in free text (box 1).

There was repeated identification of the huge time and planning burden inherent in this process and much focus on the difficulties of communication with patients in terms of timing, sensitivity and limitations of such discussions in the face of disease progression and in the absence of any prior advance decisions or planning.

“The problem of course lies with the amount of information given to the patients at commencing NIV... We now have large multidisciplinary team meetings involving the family and patient if they want to plan the terminal phase and weaning.”

The need for NIV withdrawal to be a multidisciplinary team (MDT) decision was a recurrent theme, with many commenting on the inherent challenges faced in terms of the need to support all involved, including the patient, family and staff. Managing conflicts that arise from differences in opinions within the MDT were also emphasised as practical challenges.

“I personally find it most challenging when there is difficulty reaching consensus within the clinical team...”

### Box 1 Practical issues in non-invasive ventilation (NIV) withdrawal identified by respondents

“The practical issue of whether sedating before withdrawing is the correct thing to do—what if they die before you withdraw? The practical issue of whether to wean the NIV or just stop it?”

“Practically challenging decision of who actually removes the mask, who administers any sedative, who judges the correct dose to give and decision whether to give sedation IV.”

“With lack of guidance it has been difficult to ensure you are correctly managing the procedure without causing the patient too much distress.”

### Box 2 Ethical challenges identified

“Creates significant anxieties among families and staff when a patient requests to withdraw non-invasive ventilation. Requires a large investment of time to discuss through the ethical issues with each member of staff.”

“Factoring in the wishes of the patient re timeliness of withdrawal. ... being clear about the distinction between reaching natural end of life using deep sedation and expediting death.”

“...I think I might find this more difficult if it was being requested at home and in my mind seemed premature.”

### Ethical challenges

Ethical challenges were perhaps seen as less than the practical and emotional challenges but were still considerable.

The key themes related to the timing and appropriateness of withdrawal, the need for intentions to be clear to all and the time taken to discuss ethical issues with staff (box 2), and the issues related to capacity and ADRTs (box 3).

While some clearly stated NIV withdrawal is not euthanasia, many acknowledged how they felt the process could be construed as causing the death and was potentially open to external criticism. The complexity of the ethical stance, that withdrawal of a treatment that is no longer requested by the patient is *allowing* death to occur, rather than *causing* the death, may not be fully appreciated by all involved, even with the healthcare professionals within the team. While the ethical logic is understood, the process of NIV withdrawal, for some at least, *feels* different to the withdrawal of other treatments.

“It is withdrawing a treatment that is keeping them alive and so it feels like euthanasia...”

### Box 3 Ethical issues relating to mental capacity and the experiences of using advance decision to refuse treatment (ADRT)

“...in the context of failing communication (+/– cognition), it is hard to be confident in respecting autonomy, and where capacity has been lost it is hard to be confident where best interests lie, and either way there are often conflicts which can further undermine confidence.”

“One of the ethical challenges rests in interpreting previously expressed wishes... I have found that even (seemingly) well thought out ADRT that have been extensively discussed with the medical team do not always offer the reassurance or help to the clinical team that I thought they might.”

“Ethically—need to be clear about everyone’s wishes and intentions. I know it is not different ethically from not starting a treatment but it feels different.”

### Emotional challenges

While two respondents (one who had withdrawn NIV and one who had not) were not at all challenged emotionally by the process, over half of respondents scored this as 7 or more and 20% as 9 or 10 on the 0–10 scale.

The emotional burden which was felt in terms of managing the emotions of others (patient, family and staff) throughout the process was the commonest theme to emerge. Supporting others and conflict resolution formed part of this burden (box 4).

Concerns about causing harm or distress to the patient were also common. But perhaps the most complex emotional issue was related to death being so related to an action, albeit not the intention of the action (box 5).

## DISCUSSION AND CONCLUSIONS

This survey has identified that there are considerable challenges faced by those palliative medicine doctors involved in the withdrawal of NIV at the request of a patient with MND. It appears that compared with those doctors who *have* actually been involved in withdrawal of NIV, a greater percentage of doctors that had *not* been involved in withdrawal of NIV identify that each of the practical, ethical and emotional dimensions present a level of challenge of 7 or more to them. This could mean that experience somewhat overcomes the challenge of the situation. The perceived challenges, despite experience however, remain great. Clinical and team leadership is challenged by the rarity of the event, the lack of evidence base and guidance to draw on, and the emotional impact on the doctor themselves. Perhaps another impact of the rarity is that in addition, there are few colleagues to approach who can advise and mentor ‘novices’.

There is a clear desire that discussions with patients with MND and their families concerning the benefits and challenges of using NIV and the choices that some will face need to be better integrated into care at

### Box 4 Emotional challenges for doctors in withdrawing non-invasive ventilation (NIV)

“I found dealing with the (very highly expressed) emotions of staff watching, assisting or aware of the withdrawal of NIV in a patient was more challenging for me than the emotions of the family.”

“Greatest stress is in dealing with the families rather than the patient (and sometimes with members of the wider clinical team).”

### Box 5 Emotional impact of the consequences of action

“Although very clear as to the morals and ethics of the situation, the emotional drain was enormous as we are so unused to having that much control over the timing and place of death.”

“I do find the rapid transition from living to dying aided by the removal of a life sustaining treatment and supported by respiratory and conscious level depressant drugs to be more stressful than most other areas of practice.”

“It feels like I am causing their death even though I am carrying out their wishes and it is in fact a horrible disease that is killing them. My rational brain can eventually navigate the arguments but my heart lags a long way behind.”

earlier stages of the disease. There is little detailed published information about what happens to patients using NIV as they deteriorate and die, thus making informed consent and general sharing of information with patient and families difficult.<sup>12</sup> Better development of advance care planning (ACP) processes is crucial. However, although many patients with MND can fully engage in ACP and novel methods can facilitate this, the reality with some patients and their families is that conversations about future deterioration and the grief of loss are overwhelming, avoided or traumatic. Avoidance may be from the patient, family and professionals. Thus clarity about advance wishes ‘under what circumstances would you wish to no longer continue NIV?’ can sometimes be very difficult. For others, choice preference is ‘unstable’ and can lead to acceptance or rejection of treatments thought unlikely to be wanted when planning hypothetically.<sup>30</sup> This case has resonance to the findings of the SUPPORT (Study to Understand Prognoses and Preferences for Outcomes and Risks of Treatment) study which found that outcomes often did not match preferences stated in advance of the clinical need.<sup>31</sup> In general, ACP does seem to help patients who maintain capacity, to be able to engage better in decision making when the time comes, and reduces the distress of relatives if best interest decisions need to be made when the patient loses capacity.<sup>32</sup> The outcomes of ACP, specifically in patients with MND, are yet to be explored.

### The ethical context is well rehearsed

The right of a patient with capacity who requests withdrawal of a medical treatment must, in most countries including the UK, be respected and the treatment stopped.<sup>13 14 25 33 34</sup> To continue a medical treatment which the patient has said they do not want is, in most countries, illegal. The challenges to this

black and white ethical principle in real world clinical practice are multifactorial and include: “it may be that the patient wants to stop and that request must be respected, but given that they may need assistance in stopping [they can’t move their hands to remove the mask] and want to avoid suffering [so need drugs for symptom management] does it have to be me/my service that does this?”<sup>24</sup> Like the respondents to our much larger survey, authors describe considerable time, effort and emotional energy that is needed to discuss the decision with the patient and their family, and to gain understanding and agreement in the MDT about the withdrawal rationale.

The emotional challenges appear more complex and require further investigation. The ethical literature emphasises the stressful nature of this area of clinical care and how differences in opinion and belief, despite a uniform legal context, can influence a doctor’s practice and level of distress. In our survey, many respondents reported a cognitive–emotional dissonance for themselves, and many described having to manage their own unusual emotions. The emotional burden is compounded for some by the need to both support the team and sometimes shoulder antagonism from team members, echoing findings in smaller studies.<sup>21 35</sup>

Healthcare professionals working in palliative medicine were chosen as the sample population for this scoping survey purposefully with the understanding that those working in different specialities involved in withdrawal of NIV in MND (eg, respiratory or neurology specialities) may have additional insights to add. About 13% of the APM membership responded to the survey. As the survey was electronic, it is not possible to know the number who actually ‘opened’ the survey. The survey will have had relevance only to doctors who care for patients with MND, which will include an unknown percentage of members of the APM. The response rate is thus not possible to calculate. Those that responded to the survey might however be those that have experienced more challenges compared with those that did not respond. Even so, these findings are from a sizeable group of doctors in palliative medicine in the UK.

While this survey goes well beyond any other literature, it is still a very superficial exploration of this area of practice which is worthy of deeper exploration. Why is it, as one respondent put it, that withdrawal of NIV at the request of a patient with MND “in theory, ethically, legally and practically should be straightforward, in reality is absolutely not?” Moreover, how can we mitigate the challenges to best outcomes for all concerned? Consideration of targeted support for MDT members involved in withdrawal of NIV, including education, psychological support and debriefing sessions, may all be valuable.<sup>36</sup> Withdrawal of NIV from a patient who, without it, will experience severe symptoms of breathless and feelings of

suffocation requires careful planning. The when, where, who and how need to be discussed, planned and documented in detail. The results of our survey suggest that at least in part, practical challenges could be addressed by development of guidelines on NIV withdrawal for patients with MND. Ethical challenges could be alleviated by incorporating a clear ethical statement within this guideline. We suggest also that a network of mentors may support doctors undertaking this.

### What is already known on this subject

- ▶ There is little current evidence looking at the issues faced by healthcare professionals involved in the withdrawal of non-invasive ventilation at the request of patients with motor neurone disease.
- ▶ Some case reports identify this as an area of considerable complexity which may cause controversy within the healthcare team.

### What this study adds

- ▶ Those that responded identified a very high level of ethical, practical and emotional challenges compounded by the rarity of the event, the lack of evidence base and guidance to draw on, and the emotional impact on the doctors themselves.
- ▶ The study identifies the need for a better understanding of the difficulties in order to inform guidance on the subject and hence improve patient, carer and professional experiences.

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**Contributors** CRH, DO and CF devised the questionnaire, CRH led the analysis of the results and DO and CF contributed to this. CF led the writing of the paper; both CRH and DO have commented on drafts and agreed the final content.

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**Ethics approval** The opinion of the South East Research Ethics Committee was sought and the chair advised that NRES ethics approval was not required as the study constituted a service evaluation.

**Competing interests** DO was a member of the guideline development group for the NICE clinical guideline on NIV in MND Guideline 105.<sup>3</sup>

**Provenance and peer review** Not commissioned; externally peer reviewed.

**Data sharing statement** The paper includes a summary of quantitative and qualitative findings. All raw data are available to be shared if appropriately requested through contact with the corresponding author.

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