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### Citation for published version

Faull, Christina and Oliver, David J. (2016) Withdrawal of ventilation at the request of a patient with motor neurone disease: guidance for professionals. *BMJ Supportive and Palliative Care*, 6 . pp. 144-146. ISSN 2045-435X.

### DOI

<https://doi.org/10.1136/bmjspcare-2016-001139>

### Link to record in KAR

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Author's Accepted Manuscript

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# Withdrawal of ventilation at the request of a patient with motor neurone disease: guidance for professionals

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The Association for Palliative Medicine of Great Britain and Ireland (APM) has recently published Guidance for Professionals to support their practice in a very challenging area of care (1). The Guidance has been endorsed by the Motor Neurone Disease Association, the Royal College of Nursing, the Royal College of Physicians of London, the Royal College of General Practitioners and Hospice UK, and is consistent with General Medical Council (GMC) standards of good practice (2) and is compatible with coronial law and principles. It has resulted from original research from Faull *et al* (3) in Leicester which explored the experiences of families and healthcare professionals and found that while there were examples of good practice, there was considerable variation in care and evidence of poor outcomes for patients, families and the professionals involved (4,5).

Motor neurone disease (MND) is a fatal neurodegenerative disease. Although there is variation in the way MND first affects people and the pattern and speed of progression of muscle weakness, at some point almost all patients will have weakness of respiratory muscles. The most frequent cause of death is respiratory failure secondary to impairment of the respiratory musculature, usually within a few years of onset of the illness.

Non-invasive-assisted ventilation (NIV) is a medical treatment that can improve quality of life, symptoms and survival in selected patients (6). The guidance from the National Institute for Health and Care Excellence (NICE) supports its use (7,8) and the use of NIV by patients with MND has increased considerably in this century (9). Undoubtedly, NIV has many benefits for patients and perhaps the majority of patients with MND choose to at least try NIV

Most patients who do find NIV valuable use NIV for discrete periods of time, most often at night only. However, some patients use NIV for much longer periods of the day to relieve their symptoms and a small number become very dependent, unable to tolerate even a few minutes without it (such as for cleaning teeth or drinking). It is not possible to predict which patients might follow this route when treatment is started.

A very small number of patients with MND may later choose assisted ventilation via tracheostomy (TV) if NIV fails to provide sufficient support. Patients with MND who use TV have, however, more usually had this initiated in an unplanned way when presenting in crisis. The number of patients on TV is unknown but appears to be increasing and is likely to increase further with time. The majority of patients on TV will progress to use this 24 hours a day and some, but not all, will be unable to make any respiratory effort themselves.

Patient decision-making around starting NIV is complex (10). Patients may elect to start NIV for a range of reasons, although an improvement in quality of life is usually of paramount importance. The ability to control discontinuation of assisted ventilation can, for some patients, be a crucial factor for the patient making the decision about starting the treatment. It is of great importance that the patient considering NIV is aware that they can discontinue NIV at any stage in the future if this is their wish. The 2016 NICE Guidance has made recommendations for stopping non-invasive ventilation, including the explanation before starting NIV that it may be stopped at any time and that there should be opportunities for the patient's wishes for continuing or withdrawing NIV to be discussed.

For the majority of patients, assisted ventilation does not complicate the dying process; if its benefit has been lost, then those using NIV only at night may simply choose not to put it back on. For others, assisted ventilation may continue to provide benefit throughout the dying process. However, a very small but potentially increasing number of patients who are dependent on NIV, and some on TV, request that the assisted ventilation is withdrawn because of deterioration in their quality of life due to disease progression. Without the ventilator, these patients are likely to develop acute and severe breathlessness, so the process of withdrawal needs to be managed in a planned and pro- active way to ensure that they receive appropriate symptom management and that unnecessary dis- tress is avoided.

A patient who is ventilator dependent and who decides that they no longer wish to have assisted ventilation has made a difficult decision. This life-ending decision may evolve over time, but often patient decisions around treatment withdrawal arise in the setting of a clinical deterioration, either secondary to an acute problem such as infection or in the setting of a more gradual decline in function that leads to a persistently unacceptable quality of life. A decreasing ability to communicate effectively may play a significant role in decision-making. Some patients may make a written statement or an Advance Decision to Refuse Treatment with respect to withdrawal in advance of their losing the ability to communicate or losing capacity for another reason. Others may appoint an attorney for decisions about life-sustaining treatments.

Evidence suggests that too few patients know about their potential choices or are asked about their views of continuing assisted ventilation. There is a clear need for more information sharing and improvement in facilitated decision-making. This information and discussion should occur before starting NIV and throughout the disease progression, so that decisions can be made with full involvement of the patient and those close to them (8).

Professionals have said that providing the care for a ventilator- dependent patient who has asked for assisted ventilation to be withdrawn is practically and emotionally challenging and that a lack of guidance on practical aspects of withdrawal, poor advance care planning, lack of experience and the need to support all involved in order to prevent conflict were significant factors in the impact of this care on themselves and others. Additionally, although the ethics and legality are, in theory, very clear, in practice many voiced considerable uncertainty as to what constitutes ethical and legal defensibility in these scenarios.

The APM Guidance, developed by a multiprofessional group, identifies five standards for care and the processes that will support achievement of these (Table 1). The Guidance also calls for continued gathering of data and outcomes seeking submission of a defined data set by those who undertake this care. As this area of care is rare most practitioners have very limited experience to draw on and the Guidance signposts to support from colleagues with experience via a list held by the APM secretariat.

## References

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- 4 Phelps K, Regen EL, Oliver D, *et al*. Withdrawal of ventilation at the patient's request in MND: a retrospective exploration of the ethical and legal issues that have arisen for doctors in the UK. *BMJ Support Palliat Care* Published Online First: 11 Sep2015. doi: 10.1136/bmjspcare-2014-000826
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- 6 Bourke SC, Tomlinson M, Williams TL, *et al*. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial'. *Lancet Neurol* 2006;5:140–7.
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- 9 O'Neill CL, Williams TL, Peel ET, *et al*. Non-invasive ventilation in motor neuron disease: an update of current UK practice. *J Neurol Neurosurg Psychiatry* 2012;83:371–6.
- 10 Martin NH, Landau S, Janssen A, *et al*. Psychological as well as illness factors influence acceptance of non-invasive ventilation (NIV) and gastrostomy in amyotrophic lateral sclerosis (ALS): a prospective population study. *Amyotroph Lateral Scler Frontotemporal Degener* 2014;15:376–87.

Table 1 Summary of the guidance

Timing	Standard	Process to address standard(s)
When starting assisted	<p><i>Standard 1</i></p> <p>A patient should be made aware that assisted ventilation is a form of treatment and they can choose to stop it at any time.</p> <p>They should be in no doubt that this is legal and that healthcare teams will support them</p>	<p>Inform patients that they can choose to stop the treatment at any time, that it is entirely their right and legal and that their healthcare team will manage their symptoms in a different way</p> <p>Offer patients and, with due regard for confidentiality, families the opportunity to discuss future scenarios when assisted ventilation is being considered</p> <p>Promote the concept of advance care planning, and discussion of wishes and values with patients who use assisted ventilation, especially those who have lost one modality of communication.</p> <p>Assess and discuss capacity for the decision about treatment and its continuation.</p>
Withdrawal of assisted Ventilation	<p><i>Standard 2</i></p> <p>Senior clinicians should validate the patient's decision and lead the withdrawal</p> <p><i>Standard 3</i></p> <p>Withdrawal should be undertaken within a reasonable timeframe after a validated request.</p> <p><i>Standard 4</i></p> <p>Symptoms of breathlessness and distress should be anticipated and effectively managed</p>	<p>Affirm the decision by assessing the patient's capacity or validity and applicability of an advance decision to refuse treatment (ADRT) and that this is a settled view; allowing a period of time for discussion and reflection between the initial conversation and the patient's final decision</p> <p>Planning, coordination and communication are vital tasks</p> <p>Discuss with the patient and family when, where and how withdrawal will happen, including the potential for living for some hours without the ventilator and occasionally longer.</p> <p>Discuss with the professionals when, where and how withdrawal will happen; identify key people and their roles. Ensure members of the team understand the ethical principles and the legal position.</p> <p>Make a plan for symptom management. Key decisions are:</p> <p>Does the patient require <i>sedation</i> before assisted ventilation withdrawal: ventilator-dependent</p>

patients, using >16 hours a day; very short periods off ventilator before distress.

OR

Does the patient require *augmented symptom control*: can patient manage some hours off assisted ventilation?

What drugs, doses, route?

Who will prescribe and administer?

Who will manage the ventilator and how will settings be adjusted and mask/tubing removed?

Administer anticipatory medication, titrating opioids and benzodiazepine to manage symptoms

For those who are ventilator-dependent, assess effectiveness of symptom management by reducing or stopping assisted ventilation for a few minutes before full removal.

Continue to titrate opioids and benzodiazepine to manage symptoms.

After death

*Standard 5*

After the patient's death, family members should have appropriate support and opportunities to discuss the events with the professionals involved

Consider the needs of family members and professionals after death

Plan who will provide support to family members  
Debrief for professionals/significant event analysis.

Submit data set and share key learning.