

# Withdrawal of Assisted Ventilation at the Request of a Patient with Motor Neurone Disease

## Guidance for Professionals

### Association for Palliative Medicine of Great Britain and Ireland

November 2015



Association for  
Palliative Medicine  
Of Great Britain and Ireland

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Feedback on this document is welcomed and should be sent to the APM [becki@compleat-online.co.uk](mailto:becki@compleat-online.co.uk)

#### Version and review date

Version 1.0 November 2015 | Date for review October 2017

## Acknowledgments

The Association for Palliative Medicine would like to thank all those who contributed to this Guidance. This includes many professionals who have given time and expertise and shared their experiences so openly in order to improve care for patients and the outcomes for their families. As this work draws substantially on an interview study with 17 family members and 50 health professionals, we should also like to acknowledge their enormous contribution to the depth and breadth of this Guidance. Only by exploring what has happened have we been able to understand what is needed and to implement change.

More details of this project may be found on the LOROS hospice website:

<http://www.loros.co.uk/education-training-research/research/exploring-mnd-experiences/>

This work was funded by LOROS the Leicester, Leicestershire and Rutland Hospice and the Motor Neurone Disease Association.

The guidance group is grateful for the review and helpful input of Dr Gemma Andrew, Medical Adviser to the Coroner's Service of Northern Ireland, Fiona Wilcox, medico-legal secretary for the Coroners' Society of England and Wales, Dr Michael Devlin, Head of Professional Standards and Liaison the Medical Defence Union.

The development of the Guidance was supported by a grant from the Motor Neurone Disease Association.

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## Endorsements

The Education and Standards Directorate at the GMC have advised us that this guidance is consistent with the standards of good practice set out in their guidance on *Treatment and Care towards the End of Life*.

The Guidance has been reviewed by the medico-legal secretary of the Coroners' Society of England and Wales for compatibility with coronial law and principles.



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## Guidance summary and standards for care

The purpose of this Guidance is to support professionals in working with the small number of patients with motor neurone disease (MND) who are dependent on their ventilator (use it more than 16 hours a day) and ask that this assisted ventilation be withdrawn.

The use of non-invasive assisted ventilation (NIV) improves quality of life and survival in selected patients with respiratory failure due to MND. For the majority of such patients, NIV 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, NIV may continue to provide benefit throughout life and during the dying process.

However, a minority of patients with MND who are ventilator-dependent request that the assisted ventilation is withdrawn because the burdens outweigh the benefits for them. These patients are likely to develop acute and severe breathlessness without the ventilator, so the process of withdrawal needs to be managed in a planned and proactive way to ensure that they receive appropriate symptom management and that unnecessary distress is avoided.

There are degrees of ventilator dependence. Some patients will be unable to tolerate even a few minutes without assisted ventilation and others will be able to tolerate several hours. This variability requires an individualised plan of care.

There are a number of principles that underpin this Guidance and the standards for care. These principles are:

- The decision to discontinue assisted ventilation is a unique journey for every patient and their family<sup>§</sup>.
- For a patient dying from MND, it is their legal right to decide to refuse assisted ventilation, and the duty of care of professionals to manage the physical and emotional impact of this decision on the patient and family members.
- Communication with the patient, the family and between the professionals involved is of fundamental importance in achieving sensitive, safe and effective care.
- Teamwork is key to achieving best outcomes for the patient, and requires senior clinical leadership.
- The need for psychological support for the patient, the family and for the professional team should be anticipated and planned for.
- The principles for the management of symptoms are generalisable but the precise methodology requires individual tailoring to the patient.
- As this is a rare area of care, with very little published evidence base, there is a need for ongoing evaluation of methods and outcomes.

Standards for the care of a patient and their family before, during and after withdrawal of assisted ventilation and the processes that support this are summarised in the table below.

This document is structured to provide concise guidance for delivery of safe and effective care, with extensive supporting information included as appendices.

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<sup>§</sup>In this Guidance the use of the word 'family' is inclusive of those close to the patient as well as those actually related.

## Summary of the Guidance

Timing	Standard	Process to address standard(s)
When commencing assisted ventilation and throughout care	<p><b>Standard 1</b> 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><b>Page 14</b> 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><b>Standard 2</b> Senior clinicians should validate the patient's decision and lead the withdrawal.</p> <p><b>Standard 3</b> Withdrawal should be undertaken within a reasonable timeframe after a validated request</p>	<p><b>Page 14</b> 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, co-ordination and communication are vital tasks.</p> <p><b>Page 15</b> 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><b>Standard 4</b> Symptoms of breathlessness and distress should be anticipated and effectively managed</p>	<p><b>Page 18</b> Make a plan for symptom management. Key decisions are: -Does the patient require <b>sedation before</b> assisted ventilation withdrawal: ventilator-dependent patients, using &gt;16 hours a day; very short periods off ventilator before distress. Or - Does the patient require <b>augmented symptom control</b> : patient can 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 <u>before</u> full removal.  Continue to titrate opioids and benzodiazepine to manage symptoms.</p>
After death	<p><b>Standard 5</b> After the patient's death, family members should have appropriate support and opportunities to discuss the events with the professionals involved.</p>	<p><b>Page 26</b> Consider the needs of family 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.</p>

A list has been compiled of senior healthcare professionals who have experience in withdrawing assisted ventilation and who would be willing to guide and support others undertaking this. The list is held by the Secretariat of the Association for Palliative Medicine (becki@compleat-online.co.uk).

## Why this Guidance is necessary

A patient who is ventilator-dependent and who decides that they no longer wish to have assisted ventilation has come to a momentous decision. This life-ending decision evolves over time and is incredibly hard to make.

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. 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 recurrent themes.

Additionally, although the ethics and legality are, in theory, very clear, in practice many colleagues voice considerable uncertainty as to what constitutes ethical and legal defensibility in these scenarios.

Although there are some examples of good experiences, families tell us that the care has often fallen short of what they and the patient needed. In addition, they have often had specific needs which were unmet in bereavement especially as they may not have been able to share what had happened with close friends.

An increasing number of people with MND are using assisted ventilation to manage their symptoms, improve quality of life and, for many, prolong their life. Most who use non-invasive assisted ventilation (NIV) discontinue it themselves at some point, perhaps having gradually reduced its use as they feel it of less benefit as the disease progresses. However, some continue to use it until they die. A proportion of patients who use NIV become totally dependent on it, as are the much smaller number of patients who use continuous tracheostomy-delivered assisted ventilation (TV).

A small number of these ventilator-dependent patients make an elective decision to stop using assisted ventilation. Patient decisions around treatment withdrawal tend to 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 directive 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.

The aim of the Guidance is first and foremost to improve the care of patients and families. The application of the Guidance should also lessen the emotional impact on professionals. We recognise that whilst primarily aimed at professionals some of the content may also be useful to patients, families and others in supporting people with MND.

Through ongoing collection of a minimum dataset described in this Guidance (Appendix 6), it is intended that practice can continue to be evaluated and refined in order to strive for the best possible outcomes and experiences for patients, families and professionals.

## Association for Palliative Medicine Position Statement November 2015

### Withdrawal of ventilatory support at the request of an adult patient with neuro-muscular disease

This statement intends to set out the legal and ethical position for the care of patients with neuro-muscular conditions in the UK who request that their ventilatory support be withdrawn. While the ethical principles are generic and applicable across the UK, the law in relation to mental capacity differs between England and Wales combined, Northern Ireland and Scotland (see Appendix 2). For simplicity the rest of this document draws on the 2005 Mental Capacity Act (MCA) for England and Wales.

1. In UK law, a refusal of a medical treatment by a patient who has capacity for that decision, must be respected and complied with, even if to comply with this refusal could lead to significant harm to the patient, including to their death. To continue medical treatments that a patient does not want is to give treatment without consent, and legally constitutes a criminal offence of battery or a tort in civil law, justifying financial compensation.
2. Assisted ventilation, whether invasive and delivered through a tracheal tube, or non-invasive and delivered by a mask or other equipment, is a medical treatment.
3. A patient with capacity to make such a decision may either refuse assisted ventilation or ask that it be withdrawn.
4. A patient with capacity may also make an advance decision to refuse treatment (ADRT) to be implemented at a future point when capacity is lost and the specified circumstances for the refusal become applicable.
5. Whilst the timing of death will be influenced by the withdrawal of ventilation in these circumstances, the cause of death from a medical perspective remains the advanced neurological disease, and the classification of the death should be natural causes\*\* for the purposes of issuing a medical certificate of cause of death and subsequent registration of the death by the next of kin. Such a certificate may read for example:  
1a ventilatory failure (due to) 1b advanced motor neurone disease.
6. When a patient with capacity has decided that the burdens of continued medical treatment outweigh the benefits, this is distinct in law from a decision to foreshorten their life by suicide or 'self-neglect'.
7. Withdrawing a medical treatment that a patient with capacity no longer wants, even if this is considered life-sustaining, is not assisted suicide.
8. Withdrawing a medical treatment from a patient who no longer has capacity, but who while having capacity made an advance decision to refuse treatment (ADRT) which is specific in this regard and valid for these particular circumstances, is not euthanasia even if the medical treatment is life-sustaining.
9. Withdrawing a medical treatment from a patient who no longer has capacity, on the advice or request from an appointed attorney, including decisions on life-sustaining medical

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\*\*In neurological conditions where the origins of the disability are 'unnatural', such as ventilator-dependent high traumatic spinal cord injury, the death is reportable to the coroner



treatment, and where on multidisciplinary review this request meets 'best interests' criteria, is not euthanasia even if the medical treatment is life-sustaining.

10. Withdrawing a medical treatment from a patient who no longer has capacity but who has not made an ADRT or appointed an attorney, is a conventional 'best interests' determination; the principles of which are set out within the Mental Capacity Act 2005 and refined within more recent case law.
11. Patients and clinicians should openly discuss their thoughts and concerns about assisted ventilation and quality of life, and the circumstances in which a life sustained by ventilatory support would become intolerable or unacceptable. These discussions involving the patient, their family (with due regard for confidentiality) and the multidisciplinary team preferably should begin before assisted ventilation starts and continue throughout the duration of the illness.
12. Discussion of factors leading to the decision to stop assisted ventilation should be open, without coercion and thorough, seeking to identify any potential for alternative decisions and to minimise the impact of such a decision on family members. Ideally such discussion should be with the individual patient, family and healthcare team members, with these key people together.
13. Assessment of capacity to make the decision to stop ventilatory support is mandatory. As a matter of routine it should be a practitioner familiar with the issues who is assessing capacity for decision making on those issues. Given the challenges in such decisions, and in the enactment of Advance Decisions to Refuse Treatment, it may sometimes be advisable to involve more than one appropriately trained clinician in assessing the patient's capacity, and to gather feedback from the multi-professional team and the family regarding the consistency of the patient's wishes. Rarely this may require additional expertise such as that of a psychiatrist to determine whether there is an identifiable and treatable mental-health disorder compromising capacity.
14. The clinical conditions where ventilatory support is required to sustain life also involve conditions where patients often cannot physically withdraw assisted ventilation themselves and so it will need to be withdrawn by the clinical team.
15. Withdrawing assisted ventilation may lead to distressing symptoms that require anticipatory and timely treatment with appropriate doses of medications such as sedatives and opioids targeted at relieving these symptoms. As with all good practice in palliative care, the intent must be solely to avoid or ameliorate symptoms of discomfort or distress. Relieving a patient of discomfort and distress is a fundamental medical responsibility and is not a modifier of the cause of death as set out above.
16. This area of care is challenging and requires excellence in multidisciplinary working and clinical leadership. Input from specialist palliative care will be helpful and support for members of the team is important.
17. The GMC guidance *Treatment and Care towards the End of Life: Good Practice in Decision Making* (2010) provides more detail including how to conduct this decision making in the context of conflict, disagreement and questions with respect to mental capacity and in particular the value of gaining a second opinion in these cases.

## Part 1: The context

### An overview of assisted ventilation and withdrawal in MND

MND is a fatal, adult onset, neurodegenerative disease. Patients vary in the way MND first affects them, the speed of progression and the pattern of progressive weakness of muscles, but at some point almost all have weakness of respiratory muscles. The most frequent cause of death is respiratory failure secondary to impairment of the respiratory musculature, usually within 3 years of onset.

Non-invasive assisted ventilation (NIV) is a medical treatment that can improve quality of life, symptoms and survival in selected patients. Guidance from the National Institute for Health and Care Excellence (NICE) supports its use. Undoubtedly, NIV has many benefits for patients and the majority of patients choose to at least try NIV.

A very small number of patients may later choose assisted ventilation via tracheostomy (TV) if NIV fails, and some have TV initiated in an unplanned way when presenting in crisis. In the UK the use of NIV has increased markedly over the past 10 years. The number of patients on TV is unknown but appears to be increasing and is likely to increase further with time.

Patient decision making around starting NIV is complex. 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.

Most patients use NIV for discrete periods of time, most often at night only. Some patients use NIV for much longer periods of the day and a small number become very dependent, unable to tolerate even a few minutes without it (e.g. for cleaning teeth or drinking). 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.

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, NIV may continue to provide benefit throughout the dying process. Perhaps around half of patients using NIV in the UK die having stopped it themselves (not put it back on at some point) and half die while still using NIV.

However, a very small but potentially increasing number of patients (a tentative estimate of 1–5%) 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. These patients come to a decision that the burdens outweigh the benefits for them. It is not possible to predict which patients might follow this route when treatment is commenced.

Some may make a written statement or directive with respect to withdrawal, or appoint an attorney for decisions about life-sustaining treatments, in advance of their losing the ability to communicate or losing capacity for another reason.

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 proactive way to ensure that they receive appropriate symptom management and that unnecessary distress is avoided.

## **The impact of withdrawal of assisted ventilation on patients and their families**

There is no research that informs us directly about the impact on patients themselves of thinking about and making the decision to stop assisted ventilation. However, we do have the documented views of the family members and professionals who have reflected on this in a recent interview study.

While many family members reflected positively on this time as achieving what the patient wanted and some had an experience that was 'as good as it could be', many had experiences that could be improved. These interviews reveal that:

- The role of the family in advocating for the patient and achieving their wishes can be a very heavy burden.
- There may be conflict within the family, which needs to be managed.
- There is insufficient information given to patients and families about choices and reassurances about the legal right to stop assisted ventilation.
- More opportunities to discuss concerns and options should be offered.
- Once they have expressed their choice to withdraw treatment, delays in carrying out the patient's wishes are distressing for patients and their families.
- Families need information about what will be done in the withdrawal process, how long it might take for the patient to die and any symptoms or changes they may expect to see (e.g. gasping, colour change).
- Families are frequently asked or even expected to take a role in the actual withdrawal of the ventilator, and are sometimes left alone during the process.
- Families are supported by teams who are inexperienced and sometimes unconfident. This can lead to poor outcomes which families felt could have been prevented by experienced specialist input.
- If a patient experiences distressing symptoms during withdrawal, this has a significant effect on the relatives present. Relatives' perceptions of distressing symptoms may be different to those of the healthcare professionals involved.
- Family members can feel very isolated and unable to discuss the situation before or after with their usual social support network.
- Relatives may need additional support after the withdrawal and still have questions and issues they need to discuss with the healthcare professionals involved.
- Some family members were left with feelings of guilt or shame and did not discuss the patient's death with friends for fear of being mis-judged as assisting suicide.
- Cases where the withdrawal does not proceed as expected (e.g. delays in enacting the decision, symptoms not controlled, withdrawal requiring several attempts, relative being required to play a more active role than anticipated) are more likely to leave relatives with negative feelings about the withdrawal.

## **Challenges in the withdrawal of assisted ventilation**

Interviews with health professionals and family members about their experiences in withdrawing assisted ventilation at the request of a ventilator-dependent patient with MND identified key challenges:

- This is a rare event and professionals have limited experience, many a single occurrence in their lifetime practice. Home respiratory support teams accumulate the most individual experience but their level of involvement with patients with MND is variable across the UK.
- There is considerable variation in practice in where, how and by whom this is done and variance in the outcomes for patients, families and professionals.
- The emotional stakes are very high and are frequently compounded by misunderstandings of the law and ethics and by the influence of personal beliefs.
- While a framework that enables safest and most effective care is definable there is no single 'right' methodology to address symptom management and withdraw assisted ventilation. Each patient requires care individualised according to their physical and psychosocial situation.

Ethical and moral uncertainty surrounds the withdrawal of assisted ventilation in practice. Many respondents had experienced negative reactions from healthcare professionals who were unclear of the distinction between palliation of symptoms, withdrawal of treatment and physician-assisted death. This resulted in considerable emotional impact on the professionals involved in the withdrawal and on the patients and families themselves.

## **The evidence base for symptom management during withdrawal of assisted ventilation**

There is a small volume of literature discussing the clinical and practical aspects of withdrawal of assisted ventilation in MND and a little on the withdrawal of assisted ventilation for other conscious patients. However, the optimum process for this is not established and there is considerable variation in current practice.

Providing anticipatory medication to avoid discomfort and distress is a fundamental medical responsibility and parallels the use of both local and general anaesthesia or sedation prior to invasive interventions. This is an aspect of care, however, which requires certain safeguards to ensure professional defensibility.

There is some evidence that opioids and benzodiazepines, in doses titrated to manage symptoms and distress, do not shorten life (and may paradoxically delay death) in either unconscious or conscious patients.

Removal of assisted ventilation from a ventilator-dependent patient will inevitably be followed by death. Although this is usually within hours, this is surprisingly variable. This duration is often unexpected by patients, families and the professional team.

## **The impact of withdrawal of assisted ventilation on health professionals**

Stopping something that has been keeping a person alive is a difficult situation for all concerned. The withdrawal of life-sustaining treatment from insentient patients who are dying from organ failure or brain injury (not MND) in intensive care units has an emotional impact on professional team members, and physicians consider mechanical ventilation the most difficult treatment to withdraw. 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.

The withdrawal of a ventilator appears to generate more concern than withdrawing other treatments, for example fluids, in people with advanced disease. This may be because it requires a specific act that will result in death soon after. Although the death is due to the MND it can feel that the removal of a treatment caused the death and the often short time period between treatment discontinuation and death can be challenging for all concerned. The feelings engendered by the deliberate planning of a time to withdraw treatment and thus death are magnified by concerns about being seen erroneously to be assisting dying.

Some professionals may not feel able to support the withdrawal of assisted ventilation on religious grounds. Guidance from the General Medical Council (GMC) acknowledges this but requires that professionals make sure the patient is referred to another practitioner for this care.

*79. You can withdraw from providing care if your religious, moral or other personal beliefs about providing life-prolonging treatment lead you to object to complying with: (a) a patient's decision to refuse such treatment, or (b) a decision that providing such treatment is not of overall benefit to a patient who lacks capacity to decide.*

*However, you must not do so without first ensuring that arrangements have been made for another doctor to take over your role. It is not acceptable to withdraw from a patient's care if this would leave the patient or colleagues with nowhere to turn.*

*(GMC 2010)*

The Nursing and Midwifery Council (NMC) supports this statement.

There is indication then that not only does the consideration of, or actual withdrawal of, assisted ventilation have potential for 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 needs to be anticipated and planned for.

## Part 2: Key components for safe and effective withdrawal of assisted ventilation in the context of the request of a patient who is ventilator-dependent

### 1. Sharing information and discussing choices

#### Standard 1

**A patient should be made aware that assisted ventilation is a form of treatment and that they can choose to stop it at any time. They should be in no doubt that this is legal and that healthcare teams will support them**

When ventilation becomes impaired, patients and, with due regard for confidentiality, families should be offered information (verbal and written) and the opportunity to discuss and ask questions about the benefits and burdens of ventilator support. Some discussion about potential future choices and scenarios, including ventilator dependence may also be important for patients (see Appendix 3).

The process of consent for assisted ventilation should include reassurance that it can be used for as long as it is helpful and can be stopped at any time at the patient's request. There is a case for more formalised consent for initiation of assisted ventilation.

An example of how this discussion may be introduced at initiation of assisted ventilation is:

'You can stop this treatment at any time you want to. If you are using it a lot you may want some help to manage any problems such as increased breathlessness that may occur when you stop it.'

Throughout the ongoing care the patient should be offered the opportunity to discuss their concerns, quality of life and possible future scenarios. It is very important to promote the concept of advance care planning, especially for those who have lost one modality of communication (speech or writing). Even if this discussion does not result in something as specific and prescriptive as an ADRT, a record of 'values and beliefs' can be helpful in reaching a best-interests decision in the event of loss of capacity. It also helps patients develop their views.

Patients should be reassured that if there comes a point where ventilatory support is withdrawn, that the healthcare team will aim to pre-empt any discomfort and distress and will actively treat any symptoms that arise.

The ethical and legal principles and their application to decision making are discussed in Appendix 2.

### 2. Deciding and planning the withdrawal

#### Standard 2

**Senior clinicians should validate the patient's decision and lead the withdrawal**

A senior doctor should take responsibility for validating the decision to withdraw assisted ventilation and the planning and undertaking of the withdrawal. Another senior colleague may co-ordinate the

process and the team. Communication with the patient and family members and with the broad professional team is a key component of high quality care.

Patients make settled decisions about the withdrawal of assisted ventilation over time and many factors support and influence this. The journey towards the decision to withdraw assisted ventilation and the role of professionals in this journey is discussed in detail in Appendix 3.

Although for some patients this potential decision has been known about for some time, the point at which they will make a final decision that the burdens of assisted ventilation outweigh its benefits, is unpredictable and usually one of immediacy ['I want this to stop now'] although some patients will identify a future time ['I want this to stop after Christmas'].

### **Validating the decision**

A senior doctor needs to ensure that it is a settled decision of a patient with capacity or that the advance decision is valid and applicable. Precisely what is required for this will vary from patient to patient as illustrated in the cases in Section 4 below, drawn from real patients.

With due respect for patient confidentiality the family must also have an opportunity to share information, ask questions and express any concerns. There should be discussion with the patient and family on at least two separate occasions and ideally involving two different senior healthcare professionals. If there are divergent views between the patient and their family, it is useful to obtain professional and medico-legal support and guidance.

The rationale for the decision to proceed with withdrawal and the process for the evaluation of the decision should be clearly documented. This may include:

- Who made the decision
- What evidence was considered (including consideration of validity and applicability of ADRT)
- Who was involved in discussions
- That alternative approaches are known and rejected by the patient
- That the patient knows they will die as a consequence of withdrawal
- That there is no coercion, nor is the decision driven by mistaken kindness to the family
- That this is a settled view of the patient
- Capacity assessment
- Summary of the benefits and burdens (if applicable)
- Statement of best interests (if applicable).

See Appendix 2 for additional guidance.

### **Standard 3**

#### **Withdrawal should be undertaken within a reasonable timeframe after a validated request**

When a patient has reached this momentous decision it is understandably distressing to both them and their family if actions are not taken quickly (unless the patient has identified a later date). Practically, in most part because of the need for professional availability to look after the patient in their own home, there will be gaps between the request, its validation and the withdrawal. Patients and families need to understand this and be supported in this time. It would seem reasonable that this delay is in the order of a few days at most.

## **Planning and co-ordination**

For a patient who has had open discussion about their wishes for some time, it should already be clear who will co-ordinate the process, although where the patient is to be cared for will influence this. The co-ordinator's role is to ensure all elements of the withdrawal are well-planned including: effective communication with the patient, family and across the professional team; assessment and discussion of risk and conflict; identification of roles and responsibilities; the plan and availability of the drugs and equipment for undertaking the withdrawal. The co-ordinator should also prompt the completion of the audit after the patient has died. A checklist for the elements of this role is provided in Appendix 4.

The co-ordinator may be the:

- Home assisted-ventilation specialist nurse/physiotherapist/physiologist or consultant
- MND or palliative care specialist nurse
- Consultant in palliative medicine
- Neurologist
- Intensivist
- District Nurse
- GP

## **What to plan for**

### **Who?**

- needs to be informed about the planned withdrawal (MDT, family, other)?
- will manage the ventilator?
- will administer the medication?
- will have the key role of supporting the family?
- will confirm that the patient has died and inform professional team members?
- will complete the medial certificate of cause of death

A minimum of three people are needed to be there, at least at the start of the process: one to manage the ventilator, one to manage the symptom management with medication adjustment and one to focus on supporting the family. For those patients who are likely to need rapid adjustment to symptom management (likely to be those most dependent on ventilatory support) a doctor should plan to be with the patient for the entirety of the time.

### **Where will it take place?**

- The patient's preference may or may not be feasible
- What are the anticipated challenges for the preferred place and how can they be overcome?
- Is there a difference in preference for the patient and family, and what problems does this cause for the family?

### **When will it take place?**

- Provision for professional continuity of support will need to be planned, for the potential of several hours (doctors) and up to 48 hours (nurses).
- Co-ordination of professionals' availability and family support (especially if family travelling to be there).



### **What will be done in practical terms? (see Section 3 below)**

- What drugs will be used?
- How will drugs be delivered?
- What other equipment will be required and how will it be organised? (see check list Appendix 4)
- How will mask/tube and ventilator be managed? Are there clear instructions as to how to turn the ventilator off?
- What will be done with tube/mask/equipment immediately after death?
- Has a DNACPR been completed?

### **What to discuss with patient and family**

The elements of the discussion with the patient should, with due respect for confidentiality, also be had with the family. This should ideally take place at the same time unless this is not desired by the parties concerned.

The difficulty and impact of the decision should be acknowledged, and concerns and expectations explored. Several members of the MDT, including chaplaincy, can support these discussions.

The ethical and legal position underpinning the withdrawal of assisted ventilation and the distinction between assisted death and stopping life-prolonging treatment can be important to discuss, and helps the patient and family to gain confidence in the team.

### **Where does the patient wish to be?**

Discuss different care settings and implications of each.

### **Discuss and explain the process of what will actually happen**

Take into account the patient's preferences and be guided by the patient as to what detail they would like to know:

- Timing of withdrawal – this may depend on setting, and will be dependent on professional availability. Acknowledge and attempt to minimise the distress that a delay can cause. Discuss use of this time to say goodbye.
- Symptom control – including likely symptoms, what medications will be given, how they will be given, what the objectives are including level of sedation and patients' preferences.
- Mechanics of withdrawal including who will do what and, if applicable, that the level of symptom control will be tested by stopping ventilation and restarting it, to ensure all is well before finally stopping.
- What will happen once the mask/ventilation has been removed? Acknowledge the uncertainty about rate of deterioration to death on stopping assisted ventilation. Allay expectations that this will be immediate and explain it could be some hours (and longer in exceptional circumstances).
- Address fears about distress and what would happen should distress be evident.
- Discuss what additional professional support may be needed until death occurs.

### **Who would the patient ideally like to be present?**

Advise about the healthcare professionals who will need to be present and discuss the presence of friends and family. Spiritual support and religious rituals may be of significance for the patient. The availability of a nearby room for family is important as often they need some space for 'time out'.

### **Additional points for discussion with the family**

- What, if any, role might they wish to have?
- Explain physiological changes that may occur – e.g. breathing patterns and colour changes that may occur when someone is dying.
- For a patient using TV, preferences for what to do with the tracheal tube should be explored.

It may be appropriate to discuss the practicalities of death, and arrangements after death at this time, with further discussion after the patient has died.

### **What to discuss with other professionals**

A list has been compiled of senior healthcare professionals, who have experience in withdrawing assisted ventilation, who would be willing to guide and support others undertaking this. The list is held by the Secretariat of the Association for Palliative Medicine (becki@compleat-online.co.uk).

Discussions with the MDT involved in the care of the patient can be one of the most complex and time-intensive parts of the preparatory work. It can also cause considerable tension and emotional burden, and should not be underestimated.

The lead doctor and/or the named co-ordinator should discuss with those who have been caring for the patient:

- Legal and ethical contexts
- Intent of and use of medications to manage symptoms
- The specific roles of professionals at the time of withdrawal
- That those with strongly held beliefs may withdraw from providing care.

## **3. Undertaking the withdrawal and symptom management**

### **Standard 4**

#### **Symptoms of breathlessness and distress should be anticipated and effectively managed**

Although each case will vary, the withdrawal of assisted ventilation is likely to lead to breathlessness and distress, which may be rapid in onset, and should therefore be anticipated and managed proactively.

The position at law in relation to relief of discomfort and distress remains unchanged since made explicit within the judgment of Bodkin Adams in 1957; *'if the purpose of medicine, the restoration of health, can no longer be achieved there is still much for a doctor to do, and he is entitled to do all that is proper and necessary to relieve pain and suffering, even if the measures he takes may incidentally shorten life'* (*R v Bodkin Adams* [1957] CLR 365).

### **Principles for symptom management**

The approach needs to be tailored to the individual and their circumstances. The factors that may influence the specific plan will include:

- How quickly the patient becomes distressed without assisted ventilation
- Choice of drugs
- What drugs the patient is already on

- Which route of administration the clinical team feel confident with
- Who is administering medication
- The preferences of the patient and family
- The ease of venous cannulation.

These principles will be considered in detail below. Since it is recognised there are a number of potential approaches to symptom management, case examples are given in Section 4 to demonstrate how these principles may be reflected in real-world practice.

Best outcomes result from consideration of patients in two groups related to level of ventilator dependence:

### **Group S: Sedation**

**This group refers to those patients who are highly dependent on assisted ventilation and become very breathless or distressed within minutes of not having this in place. These patients will require sedation before assisted ventilation is stopped.**

It is important in this group that the level of sedation is adequate before the ventilator is removed in order to prevent distress. Bolus medication will therefore be required in this group at the start of the process.

Before assisted ventilation is completely removed the adequacy of sedation for this patient group should be assessed by reducing or stopping ventilation for a short time, reinstating it with adjustment of medication as necessary. This can be done by changing the ventilator settings, by turning the ventilator off, or by removing the mask/interface.

The degree of sedation required for effective management of symptoms for these patients is that which achieves a reduced conscious level with no response to voice or painful stimulus and on the 'test' reduction of the assisted ventilation, no symptoms are precipitated.

Further medication is then titrated in response to any symptoms that may arise. It may occasionally be appropriate to manage symptom distress by temporarily putting the assisted ventilation back in place while medication changes can take place and an adequate level of sedation is achieved.

### **Group ASC: Augmented Symptom Control**

**This group refers to those patients who can tolerate longer periods of time without assisted ventilation will develop symptoms after a longer period of time and will require augmented symptom control.**

In this group, sedation to a level of lack of response to voice or pain may not be required before the ventilator is removed but effective, anticipatory management of breathlessness or distress remains paramount. Most patients require medication that allows them to remain calm and mildly drowsy.

## **Medication**

### Route of administration

Both subcutaneous (SC) and intravenous (IV) administration have been used very successfully. Arguably the IV route gives the most control and responsiveness to distress. Whilst the IV route is commonly used in intensive care or hospital practice there is much less use of the IV route for any purpose in hospices or in the community. In these settings subcutaneous (SC) administration of drugs is the norm.

It is important that a plan for symptom management is developed that focuses on patient choices (especially for place of care), allows professionals to feel confident and comfortable and takes into account speed of effect of medication.

Real-world case examples of both routes of administration are given in Section 4.

Buccal and intranasal administration are relatively common in paediatric practice but less so in adults. The enteral route is the most individually variable in speed and bioavailability and thus not recommended.

### Agents

Opioid and benzodiazepine medication should be used to manage breathlessness and distress respectively and are usually combined to achieve symptom control.

Opioid: morphine and diamorphine are most commonly used but others would work as effectively and should be chosen on the basis of familiarity for the professional administering the drug.

Benzodiazepine: midazolam has the most flexibility in routes of administration but lorazepam is an alternative.

Levomepromazine may be a useful second-line sedative, especially if a patient is benzodiazepine-tolerant or already on large doses. Suggested initial dose 25mg SC.

### Doses

The dose of medication that patients require to manage symptoms is quite variable. The doses below are appropriate starting points. It is vital to titrate medication to effective symptom management. Some patients may require high or lower doses to achieve this. The appropriate dose may vary with age and physiological resilience, and other reasons including the medications already in use for management of the patient's symptoms.

#### **To achieve sedation for opioid naïve<sup>††</sup> patients who are ventilator-dependent (Group S):**

- SC: Morphine 10mg-15mg with Midazolam 10mg-15mg stat as initial dose, assessed for effect 15-20 minutes later. Further aliquots of 5mg-10mg of both opioid and benzodiazepine titrated to level of sedation.
- IV: Morphine and, separately, midazolam each diluted 1mg-2mg/ml given by slow intravenous injection in aliquots of 1mg-5mg observing for effect and titrated to level of sedation required.

The period of time between the anticipatory administration of medication and the removal of assisted ventilation is influenced by the route of drug administration and by independent patient variables. Sufficient time must be allowed to ensure that the patient has an adequate level of sedation before assisted ventilation is withdrawn. For IV administration, this may require on average 15-30 minutes of assessment and titration of medication. For SC administration, this may be considerably longer and this should be factored into the care plan.

Before assisted ventilation is removed the adequacy of symptom management should be assessed by reducing inspiratory ventilation pressure (IPAP) by around 50%, or by temporarily switching the ventilator off, or by removing the mask/interface for a few minutes, with adjustment of medication

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<sup>††</sup> For a patient who is already receiving opioids, the dose of drugs administered should be adjusted to the 4-hour equivalent dose plus a 50% increase

as necessary. Several such tests of the adequacy of symptom management and adjustment of medication are occasionally required.

**To achieve augmented symptom control for opioid naïve patients<sup>††</sup> who are less ventilator-dependent (Group ASC):**

- Morphine 5mg-10mg SC or 2mg-5mg IV with Midazolam 5mg-10mg SC or 2mg-5mg IV.
- Repeat similar doses of opioid for breathlessness and/or benzodiazepine for distress administered in relation to symptoms.

The timing of the initial medication is related to the degree of ventilation impairment, the effect of any SC infusion already commenced in preparation for the withdrawal, and the route of administration of the drug. Most but not all patients will require a stat dose of medication either 20-30 minutes before (SC) or at the time of (IV) mask removal.

Very vigilant monitoring for early signs of distress is crucial, as there is a risk of under treating, being especially mindful that SC medication will take at least 10 minutes to have effect.

**Other aspects of symptom management**

Oxygen<sup>§§</sup>

In addition to medication approaches, oxygen (if easily available and acceptable to the patient and family) may offer some control of symptoms that may be related to hypoxia. It can be applied via a nasal cannula or via tracheostomy. However, the level of evidence is low and there is no need to obtain oxygen if this increases the complexity of the end-of-life care plan.

Position

Breathlessness may be less sitting up and become acutely severe on lying down. The position and place (bed or chair) for the patient needs to be considered as part of the plan of care and symptom management.

The ventilator

Familiarisation with the ventilator is crucial before withdrawal of assisted-ventilation takes place. It is vital to know at a minimum how to:

- turn off the machine [written instructions for this should be part of the care plan]
- turn off or adjust alarm settings.

Putting the ventilator or the mask/interface back on temporarily while adjusting medication is a key part of symptom management in the early part of withdrawing treatment in Group S patients, and may very occasionally be a useful strategy at other times in the withdrawal.

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<sup>††</sup> For a patient who is already receiving opioids, the dose of drugs administered should be adjusted to the 4-hour equivalent dose plus a 50% increase

<sup>§§</sup> Oxygen should generally be used with caution in MND patients who are breathless and not on assisted ventilation, since it may lead to hypoventilation and hypercapnia.

## 4. Case examples

These are anonymised, real-world examples to give insight into what care may look like. They are not all 'perfect', but intentionally illustrate variability in patient need, the challenges that arise and how the clinical approach needs to be responsive to this. In reality, each of the clinicians reflected on areas they would probably do differently 'next time' in the technical aspects of withdrawal, and we must continue to learn from such reflections through collection of the data in Appendix 6.

### Examples of the decision-making process

JL had early onset respiratory failure and NIV allowed him to continue live a full life for many months. He mentioned to his MND specialist nurse that he now felt the burdens outweighed the benefits and he wished to stop the assisted ventilation. A consultant in palliative medicine met him for the first time a few days later. JL repeated his views. He was assessed for capacity for the decision, depression was excluded and the rationale for his decision and alternatives discussed. JL was settled on his decision. His wife supported it and said they had discussed ceilings of care since the initial diagnosis and this was a consistent view throughout his illness. JL wanted treatment to stop as soon as possible, as continuing with it, having made the decision, was unbearable. A case conference was called for the next day with the GP, palliative care consultant, key nursing staff and family members. JL thanked everyone for their input and outlined his wishes again. A plan was agreed for withdrawal of treatment at home the next day by the GP and community nursing staff.

VE was 47 and presented with bulbar MND with respiratory failure soon after. When NIV failed he chose TV. He was able to communicate in writing. At the time of commencing TV a DNACPR decision was made with his agreement. On many occasions he was encouraged to think about plans for his assisted ventilation, especially because of the impact of repeated chest infections, each time responding 'as long as I can communicate I have life.' Further discussions about the future appeared to impact adversely on his mood. Eighteen months after TV commenced he attempted to remove his ventilation himself and was admitted to the hospice in psychological crisis. Over 2 months he developed an ADRT and decided that after Christmas (some 3 months hence) he wished his assisted ventilation to stop. His mood significantly improved because of this decision. In January he confirmed this continued wish and ventilation was withdrawn at home.

KS presented in acute respiratory failure to the medical admissions unit. He lacked capacity and was commenced on NIV. As his condition improved and he regained capacity some hours later he asked that the treatment be stopped as he did not want assisted ventilation. The consultant had not met him before and was unable to talk with other professionals who had. The family supported the patient's wish but were very distressed at the prospect of him dying. A decision was made for further discussion the following day and if KS's view remained the same then the NIV would be stopped. The next day KS restated his wish, the NIV was withdrawn and KS died.

Three days into a hospice respite admission, GH decided that this was the right time to stop ventilation. There had been open, documented discussions with the MND team and GP for some time about her likely intention to make this decision, but it was not expected that it would be during this admission. Her GP and the palliative care consultant discussed the decision and the timing with her and assessed capacity. The NIV was withdrawn 6 hours later.

PQ had lost all communication. His ADRT developed with his GP stated that he would want to stop his TV when he lost communication. The GP assessed his ADRT as valid and applicable. The palliative care consultant who had not met PQ before visited and assessed PQ and the ADRT. His wife and

family raised no concerns that PQ had changed his mind at any point since making his ADRT. Withdrawal of ventilation was planned with the family for a few days later at home.

#### **An example of anticipatory sedation using the SC route**

YB was 73 and asked for his NIV to be withdrawn at home with his wife and two children. He was distressed by the prospect of delay in waiting for a doctor to be available and the withdrawal was undertaken by a respiratory specialist nurse and community staff nurse. The GP prescribed the medication and the plan was supported by a Consultant in palliative medicine.

YB was on no medication prior to the withdrawal. Drugs were administered SC.

Time zero: Midazolam 5mg + Morphine 5mg  
30 minutes: Drowsy but eyes open. Midazolam 5mg  
45 minutes: Asleep: Test of assisted-ventilation removal led to some distress. Mask replaced and further Midazolam 5mg  
75 minutes: Deeply asleep: no reaction to test of assisted-ventilation removal. Assisted ventilation stopped  
90 minutes: Asleep but restless midazolam 5mg  
100 minutes: Rapid shallow breathing: Morphine 5mg  
115 minutes: Breathing changed to slower rate  
120 minutes: Died

#### **An example of anticipatory sedation where the patient was already on high doses of opioids and benzodiazepines**

XC was 56 and decided to stop her assisted ventilation during a respite stay at the hospice since it was no longer providing effective symptom management. Three family members were with her together with a doctor and three nursing staff. The on-call doctor registrar had known the patient for one day but was supported by the consultant who knew her well and had had many documented conversations about this intention.

She was on a SC infusion of oxycodone 50mg + midazolam 70mg/24 hours prior to the withdrawal for management of considerable breathlessness, distress and pain.

Drugs were administered SC.

Time zero: Midazolam 15mg + Levomepromazine 25mg  
30 Minutes: Patient assessed as unrousable to voice or painful stimulus. Mask removed  
35 minutes: Opened eyes: midazolam 15mg + Levomepromazine 25mg  
55 minutes: Became very pale  
60 minutes: Died

#### **An example of anticipatory sedation using the IV route**

WD was 60 and breathless within 2 minutes of NIV not being in place. She was admitted to the hospice at her request to support the discontinuation of her assisted ventilation. She was mildly breathless with NIV in place and had a SC infusion of midazolam 10mg with Morphine 5mg/24 hours.

Drugs were administered IV.

Time zero: Midazolam 10mg + Morphine 10mg

10 Minutes: Patient assessed as unrousable to voice. Ventilator IPAP pressure reduced from 10 to 6 cm H<sub>2</sub>O. No apparent distress after 15 minutes

25 minutes: Assisted ventilation removed

Remained unconscious but required further 2 x 5mg Midazolam and 1 x 5mg morphine to manage mild signs of breathlessness (nasal flaring, increase in pulse rate, increase in respiratory rate)

100 minutes: Died

#### **An example of augmented symptom control using the SC route**

ZA was 72 and asked for his NIV to be withdrawn in the hospice in his chair as lying down was uncomfortable. He could manage for a period of time off the NIV, becoming increasingly distressed over 30–40 minutes. Five of his family were present, with two doctors and a staff nurse.

Drugs were administered SC.

A SC infusion of morphine 10mg + midazolam 5mg/24 hours was commenced the night prior to the withdrawal.

Time Zero: ZA was calm and mildly drowsy. He took off his NIV and was given morphine 5mg + midazolam 5mg at the same time

30 minutes: He appeared a little restless but was not communicating verbally: morphine 5mg + midazolam 5mg

50 minutes: Mild symptoms. Drowsy but not sedated: morphine 5mg + midazolam 5mg

75 minutes: Sleepy but some mild restlessness: 6.25mg levomepromazine

80 minutes: Died

#### **An example of withdrawal of TV using the SC route for anticipatory sedation**

TN had used TV for 2 years after NIV had failed. He had requested that his TV be withdrawn at home since the burdens outweighed the benefits.

Drugs were administered SC.

Time zero: Midazolam 10mg + Morphine 10mg

Time 30: Mildly drowsy still responsive to voice. Midazolam 10mg + Morphine 10mg

Time 45: No response to voice or pain evident. Test of ventilation pressure reduction by 50% with no symptoms evident after 15 minutes. Ventilation removed.



Time 70:	Sweating and nasal flaring, eyes opened. Midazolam 10mg + morphine 10mg
Time 80:	Died

#### **An example of withdrawal of TV using the SC route for anticipatory sedation**

MF had used TV for 2 years after an emergency tracheostomy was inserted due to respiratory failure and problematic secretions. She had requested that her TV been withdrawn at home since the burdens outweighed the benefits for her.

Drugs were administered SC.

A SC infusion had been in situ for 9 months and at the time of assessment was morphine 5mg + midazolam 20mg/24 hours. Patient was awake and able to make her wishes known. The ventilator was triggering all her breaths.

Based on patient preferences it was decided to increase syringe driver with view to withdrawing the following day.

3pm	SC infusing increased to 15mg Morphine and 40mg Midazolam
8am	Patient deteriorated over about an hour became unresponsive, cold and clammy
8:30am	Stat of 5mg morphine and 5 mg midazolam given and ventilation discontinued
8:35am	Died

## **5. After withdrawal**

### **Documentation**

All the professionals involved should make appropriate documentation of:

- The decision-making process (see Section 2 above)
- A summary of the medication and other strategies for symptom management. NB contemporaneous note making is recommended as the detail can be hard to recall after the event.
- Who did what
- Patient related outcomes
- Family related outcomes
- Time of death.

The medical certificate of cause of death must be completed by a doctor who has cared for the patient in their last illness and who seen then within the last 14 days. If the patient has assisted ventilation withdrawn at home either the doctor who was present during this time should complete the certificate or the care plan must make sure that the GP has visited the patient within the 14 day timeframe.

### **Support for the family**

#### **Standard 5**

## Family members should have appropriate support and opportunities to discuss the events with the professionals involved

The family will need information and support to manage the immediate post-death processes (e.g. verification and certification of death, undertaker etc.). Families should be well prepared for this time having had the discussions described above. If there was conflict in the family this may present particular needs at this time.

The everyday lives of many families have been dominated by the many practical tasks of caring for someone with a high level of disability. The death changes everything about the structure of the day and their lives. Some additional impacts of stopping assisted ventilation are:

- The continuous sound of the ventilator is gone. The quiet can be very hard and professionals commenting on this can help.
- The social isolation that can result from deciding to stop treatment. It is hard to discuss this with friends and social contacts and sometimes even within the family. They may want to be able to discuss with someone else who has gone through this if this can be arranged.
- The family may need to revisit the decision, the legality and the processes with the professionals involved, as some find what they have seen difficult to cope with.

The clinical co-ordinator should ensure that an appropriate plan is made for follow-up support for the family. The details of this will need individual tailoring, not least because families may not live locally, but is likely to include:

- Phone contact from a senior professional involved in the care of the patient in the first few days and a few weeks later
- Signposting and phone numbers to who they could contact if they need support including MNDA connect: 08457 626262 / [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org). MNDA Scotland MNDscotland.org.uk 0141 332 3903.

### Support for professionals

Members of the MDT may need a time to debrief about the events to make sure there are no doubts about ethics and legality.

Those involved in the actual withdrawal may need to reflect on outcomes; what went well and what they would wish to improve on.

For some, being involved in an intervention that relates so closely in time to the patient dying requires more bespoke support.

## Further development of evidence-based practice and future work

The lack of published evidence and the challenges of research in this are significant impairments to quality and service improvement. We must learn from the experiences of providing this care for patients. We have developed a proforma for collection of a data set which will allow us to evaluate the effectiveness of practice and make appropriate recommendations for enhancing practice (Appendix 6).

### Suggestions for future work

The Guidance development identified three priorities for research and development:

- Longitudinal study of patient decision making. For a patient to decide in advance what treatment they would and wouldn't want in the future is hard. Evidence would suggest patients often vary from their advance plan in their actions and decisions when the real situation arises. Understanding more about this decision stability/instability in relation to decisions about assisted ventilation would be important in supporting patients.
- Improving the information given to patients and the consent process when starting NIV.
- Developing guidance on withdrawal of NIV for other patient groups.

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## **The Guidance draws on the following literature**

The guidance draws heavily on the recommendations arising from work lead by Professor Christina Faull with researchers Kay Phelps and Emma Regen. This work explored the experiences and concerns of professionals and families. The recommendations, publications abstracts and other materials can be found online at [www.loros.co.uk/education-training-research/research/exploring-mnd-experiences/](http://www.loros.co.uk/education-training-research/research/exploring-mnd-experiences/).

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## Appendices

### Appendix 1. Resources to support professionals and patients and families

A number of professionals have agreed to be contacted by others who want to discuss the withdrawal of assisted ventilation. For further information contact the Association for Palliative Medicine at [becki@compleat-online.co.uk](mailto:becki@compleat-online.co.uk) 01489 565665

Other organisations may also offer information and support:

Motor Neurone Disease Association      Karen Pearce, Director of Care.  
Karen.pearce@mndassociation.org

Royal College of Nursing      Professional Lead for Long Term Conditions 0345 7726100

#### **MNDA – Motor Neurone Disease Association - <http://www.mndassociation.org>**

This website has a wide range of useful information for patients, carers and professionals.

Information Sheet 8B: *Ventilation in motor Neurone Disease (2010)* has been included in this guidance in its entirety as a useful resource, and sign posts readers to other information that may be of value.

#### **MND Scotland – [www.MNDscotland.org.uk](http://www.MNDscotland.org.uk)**

This website has a wide range of useful information for patients, carers and professionals.

MND Factsheet 40: *Ventilation in MND (2009)*. This factsheet contains more information about TV as a choice, and encourages people to think about decisions related to stopping assisted ventilation

#### **Association for Palliative Medicine**

The Association for Palliative Medicine of Great Britain and Ireland (APM) is an association for doctors who work in or have a special interest in palliative care.

The APM's Position Statement on the '*Withdrawal of ventilatory support at the request of an adult patient with advanced neuromuscular disease*' is included in the main section of this guidance.

#### **Hospice UK – <http://www.hospiceuk.org/about-hospice-care/find-a-hospice>**

Further support and advice can be accessed via your local palliative care team. This website helps you identify your local palliative care services.

#### **Inside the Ethics Committee: *Withdrawing Treatment* (Series 7 Episode 3), BBC Radio 4**

<http://www.bbc.co.uk/programmes/b012r7jn>

This Radio 4 podcast considers the issues that may be faced by patients, carers and healthcare professionals when deciding to withdraw ventilatory support in MND.

#### **LOROS hospice research**

Exploring the experiences of families and health professionals supporting a patient with Motor Neurone Disease who requests that their ventilation be withdrawn.

<http://www.loros.co.uk/education-training-research/research/exploring-mnd-experiences/>

### **e-learning resources:**

- **My NIV** – <http://niv.mymnd.org.uk>

A guide to using non-invasive ventilation for people living with motor neurone disease developed by Sheffield MNDA care team.

- **e-ELCA** – <http://www.e-lfh.org.uk/programmes/end-of-life-care/>

This Health Education England e-training resource for end-of-life care has sessions which support development of communication skills and advance care planning for health and social-care staff.

### **Resources for professionals and patients relating to advance care planning:**

*Advance decisions to refuse treatment: A guide for health and social care professionals*, 2014.

<http://www.nhs.uk/resource-search/publications/eolc-adrt.aspx>

Advance Decisions to Refuse Treatment – <http://www.adrtnhs.co.uk>

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British Medical Association – Assessment of Mental Capacity

<http://bma.org.uk/assessingmentalcapacity>

NHS Improving Quality (2014) *Capacity, Care Planning and Advance Care Planning in Life Limiting Illness: A Guide for Health and Social Care Staff* [Online]. Available at [www.nhs.uk/resource-search/publications/eolc-ccp-and-acp.aspx](http://www.nhs.uk/resource-search/publications/eolc-ccp-and-acp.aspx) (Accessed 6 August 2015).

MND Association: Information sheet 14A. Advance Decision to Refuse Treatment

<http://www.mndassociation.org/wp-content/uploads/2015/07/14a-advance-decision-to-refuse-treatment.pdf>

MND Association: End of Life Guide

<http://www.mndassociation.org/life-with-mnd/Publications-pabmnd/end-of-life-guide>

An example ADRT form – [http://www.adrtnhs.co.uk/pdf/EoLC\\_appendix1.pdf](http://www.adrtnhs.co.uk/pdf/EoLC_appendix1.pdf)

### **Ventilator guides**

There are many types of ventilator in use across the UK. The local respiratory unit should be able to provide guidance for professionals as well as patients. Some manufacturers have web-based information.

**NIPPY machines:** [www.nippyventilator.com/download-centre/](http://www.nippyventilator.com/download-centre/)

## Appendix 2. Legal position and decision-making in practice

Whilst certain legal principles are generic and applicable across the UK, the law in relation to mental capacity differs in specifics between England and Wales combined, Scotland and Northern Ireland. However, the legal and ethical principles are the same under the devolved legislations. In Scotland the equivalent legislation is The Adults with Incapacity Act 2000. In Northern Ireland this is currently covered by common law but there is a consultation process underway for a new Mental Capacity Bill

The next sections draw on the 2005 Mental Capacity Act for England and Wales and does not cover differences of the devolved nations. Some of the key differences are outlined in sections 4 and 5 below.

### 1. Legal principles

In UK law a refusal of a medical treatment by a patient who has capacity for that decision, must be respected and complied with, even if to comply with this refusal could lead to significant harm to the patient, including to their death. To continue medical treatments that a patient does not want is to give treatment without consent and constitutes a criminal offence of battery or a tort in civil law justifying financial compensation.

Assisted ventilation, whether invasive and delivered through a tracheal tube, or non-invasive and delivered by a mask or other equipment, is a medical treatment.

A patient with capacity to make such a decision may either refuse assisted ventilation or ask that it be withdrawn.

A patient with capacity may also generate an advance decision to refuse treatment (ADRT) to be implemented at a future point when capacity is lost and the specified circumstances for the refusal become applicable.

Withdrawing a medical treatment that a patient with capacity no longer wants, even if this is considered life-sustaining such as assisted ventilation, is not 'assisted suicide'.

Withdrawing a medical treatment from a patient who no longer has capacity, but who whilst having capacity generated an ADRT which is specific in this regard and valid for these particular circumstances, is not euthanasia, murder or manslaughter even if the medical treatment is life-sustaining such as assisted ventilation.

Withdrawing a medical treatment from a patient who no longer has capacity, on the advice or request from an individual with lasting power of attorney for personal welfare (LPAHW), including decisions on life-sustaining medical treatment and where on multidisciplinary review this request meets 'best interests' criteria, is not euthanasia even if the medical treatment is life-sustaining such as assisted ventilation.

Withdrawing a medical treatment from a patient who no longer has capacity but who has not generated an ADRT or appointed an LPAHW, is a conventional 'best interests' determination, the principles of which are set out within the MCA 2005 and refined within more recent case law.

Whilst the timing of death will be influenced by the withdrawal of ventilation, in these circumstances, the cause of death from a medical perspective remains the advanced neurological disease, and the classification of the death should in most circumstances be natural causes for the purposes of registration of the death from the perspective of either coroner or the law.

This scenario, where the patient with capacity has decided that the burdens of continued medical treatment outweigh the benefits, is distinct therefore from where a patient with advanced

neurological disease has chosen to foreshorten their life either actively with a method of suicide or under the umbrella of 'self-neglect' such as by deliberate sustained starvation.

'Self-neglect' as a coronial principle and conclusion could also cover treatment refusal as seen in people with unusual or different ideas who are not trying to self-harm or kill themselves, but choosing not to follow appropriate advice, thereby creating a distinction with the request to withdraw ventilatory support in the scenario envisaged by this guidance. Suicide or self-neglect are not 'natural' causes of death and must be reported to the coroner for their investigation. Consideration will also have to be given to potential criminality if a third party is thought to have 'assisted' the patient in such circumstances.

Relieving a patient of discomfort and distress remains a fundamental medical responsibility and should not in the circumstances under consideration be interpreted as modifying the cause of death as set out above.

Providing anticipatory medication to avoid discomfort and distress is also a fundamental medical responsibility and parallels the use of both local and general anaesthesia or sedation prior to invasive interventions.

## **2. Advance care planning**

Advance care planning is a process between a person and their care providers to establish their wishes for the future. It usually takes place in the context of an illness during which a patient may lose capacity or have difficulty communicating their wishes. Discussions of this nature should be documented, regularly reviewed, communicated to others involved in the patient's care and, depending on the patient's wishes, family and carers.

A statement of wishes and preferences is a written, recorded or narrative document that states the patient's values in both clinical and non-clinical circumstances. While it is not legally binding, it can be used as an account of the person's wishes when a person loses capacity and best interests need to be established.

The MCA (2005) for England and Wales underpins advance care planning and sets the legal context for such conversations and patient directions.

## **3. Practical interpretation of the MCA 2005 in relation to decision making for a patient with and without capacity**

### ***The patient has capacity***

Capacity to make decisions is situation and time specific. Provisions to optimise the patient's ability to make decisions should be made. In MND both the cognitive and communication aspects of capacity can be affected. It is vital that communication aids are available and adequate time is allowed for such discussions, especially as patients may fatigue very quickly. To demonstrate capacity the patient needs to:

- understand the information relevant to the decision to remove/reduce the non-invasive assisted ventilation. The information should specifically include the predictability of death, but also the unpredictability of the timeframe for death and the possibility of longer-term survival
- be able to retain this information and process it
- weigh up the pros and cons and come to a decision

- communicate their decision.

A competent patient is entitled to make such a decision. However, caution is advised if the request comes out of the blue; further discussion would be required to ensure that it is the patient's considered, voluntary and settled decision.

The capacity of a patient when they generate(d) an ADRT or LPAHW should be assessed to ensure its validity.

### ***The patient has a lasting power of attorney for health and welfare (LPAHW)***

A patient may have appointed a decision making proxy to make healthcare decisions on their behalf using a health and welfare lasting power of attorney (LPAHW). A lasting power of attorney is a legal document, registered with the Office of the Public Guardian, that allows a nominated person (or persons) to make decisions on the patient's behalf should the patient lose capacity; the attorney's decision is as valid as the patient themselves making that decision.

If a patient lacks capacity to decide whether to withdraw their non-invasive assisted ventilation, this decision may be made with their attorney. The attorney can only consent to or refuse life-prolonging treatment on the person's behalf if this has been specifically stated as part of the lasting power of attorney. The attorney can only act in the patient's best interest as set out in the Mental Capacity Act; anyone with concerns about the attorney's decision can apply to the Court of Protection for a decision while continuing treatment.

### ***The Patient has an advance decision to refuse treatment (ADRT), which outlines circumstances in which they would want their assisted ventilation to be withdrawn***

If a patient who is no longer competent has an ADRT that outlines circumstances in which they would want their assisted ventilation to be withdrawn, the advance decision needs to be assessed for its validity and applicability. If it is valid, and the circumstances outlined apply, then assisted ventilation should be withdrawn in accordance with the patient's previously expressed wishes.

An ADRT is a clear set of written instructions on the declining, withholding or withdrawing of treatment in the future in the contemplation of a time when that person lacks capacity including the loss of ability to communicate. An advance decision to refuse assisted ventilation (or other life-sustaining treatment) must include the phrase 'even if my life is at risk' in order for it to be valid.

An ADRT only becomes active when the person has lost capacity for the decision at hand, otherwise there should be a normal consent process.

In the event that the patient lacks capacity but the ADRT is not valid, the advance decision can still be considered as an indication of the patient's views and wishes, and taken into account within best-interests decision.

The following check list for validity is taken from NHS Improving Quality document, *Advance decisions to refuse treatment: A guide for health and social care professionals* (2014).

You are required to maximise the person's capacity and to facilitate communication			
Question		Y/N	Answer
1	Does the person have the capacity either give consent or refuse treatment him or herself, with appropriate support where necessary		<p><b>YES:</b> The person has capacity to make the decision him or herself. The advance decision is not applicable. Ask what s/he wants to do</p> <p><b>NO:</b> Continue with checklist</p>
<b>Is the advance decision valid?</b>			
2	Has the person withdrawn the advance decision? (This can be done verbally or in writing)		<p><b>YES:</b> This is not a valid advance decision. Make sure that you have identified and recorded the evidence that the person withdrew the advance decision.</p> <p><b>NO:</b> Continue with check list</p>
3	Since making the advance decision, has the person created a lasting power of attorney (LPA) giving anybody else the authority to refuse or consent to the treatment in question?		<p><b>YES:</b> This is not a valid advance decision. The donee(s) of the LPA must give consent to or refuse the treatment. The LPA decision must be in the person's best interests.</p> <p><b>NO:</b> Continue with check list</p>
4	Has the person done anything that is clearly inconsistent with the advance decision remaining his/her fixed decision?		<p><b>YES:</b> This is not a valid advance decision. It is important to identify what the person has done, discuss this with anybody close to the person, explain why this is inconsistent with the advance decision remaining his/her fixed decision, and record your reasons.</p> <p><b>NO:</b> The advance decision is valid. Continue with the checklist.</p>
<b>Is the advance decision applicable?</b>			
5	<p>(a) Does the advance decision <b>specify</b> which treatment the person wishes to refuse?*</p> <p>(b) Is the treatment in question that specified in the advance decision?</p>		<p><b>YES:</b> to both (a) and (b): Continue with the checklist</p> <p><b>NO:</b> This is not an applicable advance decision</p>
6	If the advance decision has specified circumstances in which it is to apply, do <i>all</i> of those circumstances exist at the time		<p><b>YES:</b> Continue with the checklist</p> <p><b>NO:</b> This is not an applicable advance decision</p>

	that the decision whether to refuse treatment needs to be made?		
7	Are there reasonable grounds for believing that circumstances exist which the person did not anticipate at the time of making the advance decision and which would have affected his/her decision had s/he anticipated them?		<p><b>YES:</b> If such reasonable grounds exist, this will not be an applicable advance decision. It is important to identify the grounds, discuss this with anybody close to the person, and identify why they would have affected his/her decision had she/he anticipated them, and record your reasoning.</p> <p><b>NO:</b> Continue with the checklist</p>
<b>Life sustaining treatment</b>			
8	Is the decision both valid and applicable according to the criteria set out above?		<p><b>YES:</b> Continue with the check list</p> <p><b>NO:</b> This is not a binding advance decision to refuse the specified life sustaining treatment</p>
9	In your opinion is the treatment in question necessary to sustain the person's life?		<p><b>YES:</b> Continue with the checklist</p> <p><b>NO:</b> This is a binding advance decision to refuse the specified non-life-sustaining treatment. It must be respected and followed.</p>
10	Does the advance decision contain a statement that it is to apply even if the person's life is at risk?		<p><b>YES:</b> Continue with the checklist</p> <p><b>NO:</b> This is not a binding advance decision to refuse the specified life-sustaining treatment.</p>
11	Is the advance decision: <ul style="list-style-type: none"> <li>• In writing AND</li> <li>• Signed by the person making it or by somebody else on his behalf and at his direction AND</li> <li>• Signed by a witness responsible for witnessing the signature, not the decision</li> </ul>		<p><b>YES TO ALL:</b> This is a binding advance decision to refuse the specified life-sustaining treatment. It must be respected and followed.</p> <p><b>NO TO ANY:</b> This is not a binding advance decision to refuse the specified life-sustaining treatment</p>

\*NB It is possible to use the layman's language to specify both treatment and circumstances

***A best-interests decision is made on behalf of an incompetent patient***

If a patient is dying and lacks capacity, despite all measures to maximise capacity having been made, it may be appropriate to give consideration to discontinuing their non-invasive assisted ventilation

on a 'best interests' basis'. This could be a clinical decision because of problems with the patient-ventilation synchronisation, secretion management or burdensome distress caused by some aspect of non-invasive assisted ventilation. In these cases a formal best-interest decision may need to be made.

A best-interest decision requires those making decisions on behalf of the patient to consider the things that the person would take into account if they were making the decision. It requires consideration of the benefits and burdens of continuing the intervention and any alternatives available. In order to make sure the patient's interests are best represented it often involves many people from the multidisciplinary team and the patient's relatives, friends, carers or others who can represent the views of the patients. In the event that there is no one to represent the views of a patient, an Independent Mental Capacity Advocate (IMCA) should be appointed.

The decision-making process should be documented as well as the outcome of the decision. Best-interests decisions are time and decision specific and as such should undergo review.

The doctor needs to evaluate:

- the record of discussions with the patient
- any evidence that the patient did not act in accordance with their stated wishes
- the benefit of the assisted ventilation to the patient (not just in terms of being kept alive)
- the burden of assisted ventilation for the patient.

In the event of conflict between decision makers as to what constitutes best interests for the patient, a range of measures can be implemented starting with an independent medical opinion and progressing through involvement of a clinical ethics committee and negotiation with the various parties, through to an application to the Court of Protection.

#### **4. Some aspects of the legal position with respect to capacity in Scotland**

The statute governing the treatment of adult patients without capacity is the Adults with Incapacity (Scotland) Act 2000. Section 1(6) of the 2000 Act defines an adult as someone who has attained the age of 16. The Act requires that for any intervention (including medical treatment or its withdrawal) "account shall be taken of" the patient's present and past wishes as well as any welfare attorney that might have been appointed.

The Adults with Incapacity Act (2000) does not specifically include legislation with respect to ADRT but this should be considered as legally binding under case law, providing the following principles are fulfilled:

- consent at the time of writing was valid
- the circumstances described are applicable to those that then arise
- the ADRT is current, signed and witnessed

#### **5. Some aspects of the legal position with respect to capacity in Northern Ireland**

The Mental Capacity Bill is making its way through the Legislative Assembly at Stormont, in Belfast. It is due to go to committee stage on 23 September 2015. The Bill does not specifically legislate on advance decisions (as the Mental Capacity Act 2005 has done in England and Wales), although it is clear in clause seven that account should be taken by the decision maker of the patient's past and present wishes, including "any relevant written statement" made by them. As the Bill will apply to persons aged 16 or more, it is clear that when signed into law the situation in



Northern Ireland will be very similar to Scotland, in that doctors will be required to take into account the wishes of the patient when they were aged 16 or 17. The Bill combines mental capacity and mental health legislation.

The absence of a statutory basis for an advance decision means that the common law position will apply. The common law requires that to be valid an advance decision the person must have been aged 18 or over and been capacitous at the time it was made.

### **Appendix 3. The journey towards a decision to withdraw assisted ventilation**

Patients make settled decisions about the withdrawal of assisted ventilation over time and many factors support and influence this. Key to such decision making is the availability of timely and accurate information for the patient. This requires the patient to have the necessary facts, the opportunity to ask questions and a skilled professional to enquire and prompt thinking about future potential scenarios.

Some participants in the exploratory research work that underpins this Guidance reported that patients did not realise that they could choose to stop assisted ventilation and receive symptom management. They reported that patients had felt that their only options to end treatment were suicide or assisted suicide in Switzerland. These thoughts and the distress that they caused were apparently unknown to the professional team caring for them. Many patients do not raise this themselves.

Whilst most patients want to continue their non-invasive assisted ventilation until they die, professionals need to proactively and sensitively enquire about their thinking about the tolerability of their situation now and in the future.

#### **1. Commencing assisted ventilation**

When ventilation becomes impaired, patients and families should be offered information (verbal and written) and the opportunity to discuss and ask questions about the benefits and burdens of ventilator support and potential future choices and scenarios including ventilator dependence and withdrawal.

Patients and their families are, in general, insufficiently informed about the benefits and burdens of assisted ventilation and those who have discussed their experiences of withdrawal say they would have liked much more information at the time of starting about possible future scenarios and choices.

Many patients feel much better very quickly after commencing NIV because of improved sleep and other symptoms such as headache and fatigue. However, commencing NIV is challenging and the perceived burdens of treatment can outweigh benefits in the first days and weeks until the patient becomes comfortable with the ventilator. Many patients require very active and positive support from professionals in these first few weeks.

Discussing these challenges, how they can be overcome and helping people get through them is vital in helping them achieve longer-term wishes of improved quality of life and increased survival. A problem-orientated check list for such discussions is shown in Table 1. A useful resource for patients is *myNIV* available <http://niv.mymnd.org.uk>

Future scenarios that are useful to touch on include:

- Not everyone gets on with NIV. It is not a treatment you have to have. It aims to improve your quality of life but can also lengthen your life.
- Many patients only use NIV at night but some, at some point, use it in the day and a few may come to use it all the time.
- Many patients who use NIV stop it themselves (don't put it on) when they no longer feel it is helping them. They may need medications at some point to help manage any symptoms of breathlessness.

- Some patients may choose to use NIV until they die because it helps their breathing. They may need medications as well at some point to help manage any symptoms of breathlessness.
- A very few patients choose to have long-term assisted ventilation by a tube into the lungs because their NIV is not sufficient for them. This may be an elective procedure after discussion with their home ventilation team or after an acute intercurrent illness and invasive ventilation on an intensive care unit.
- A small number of patients who are very dependent on assisted ventilation may ask that it be stopped. This is their right, it is legal and it is not assisted suicide, but it needs to be thought through and planned carefully.

Providing information about future scenarios and end-of-life choices whilst providing positive support to patients to get them through the initial hurdles is a challenge that will require both an advanced level of skill in communication and a team approach.

It is unlikely to be appropriate to have a detailed discussion about withdrawal of NIV leading to death at this time; but clearly there are some benefits in beginning such discussions at a stage of the illness before fatigue and communication difficulties increase.

Palliative medicine doctors are skilled in discussing end-of-life choices with patients and families. If they are not already involved, consideration should be given to introducing them to the patient at this stage in a patient's care, especially if it can be anticipated that a patient may make a decision to request withdrawal of treatment in the future. This can lay the foundation for the decision making and planning of that withdrawal and contributes to good symptom management.

The MNDA has written information available for patients and families (Leaflet 8b – Appendix 7).

Commencing TV is not common in the UK but appears to be an option patients are increasingly aware of and is becoming a positive choice made by some patients with MND. Discussion may take place at any stage of MND but is most often in the context of the patient becoming less well, especially with chest infection or when undergoing a planned intervention such as gastrostomy or surgery. The discussion is about agreeing key 'ceilings' for care or interventions including: would they want full active management in ICU if the need arises? Would they wish a tracheostomy if this was necessary to maintain their respiratory function?

Occasionally TV is offered as a routine planned procedure particularly if significant bulbar symptoms make NIV less effective. The commonest scenario for TV use in the UK seems to be the choice of a young patient who wishes more time with their family and for whom NIV is no longer effective enough.

The burdens of TV treatment are high, especially for families, and these and the practical elements and challenges of care need to be discussed in detail. As this is a life-sustaining treatment, patients with MND have the prospect of developing a 'locked-in' state (alive, hearing, thinking, feeling but unable to communicate). This needs to be discussed including how choices will be made about continuation of assisted ventilation in such a situation.

Table 1. Obstacles to NIV tolerance and suggested solutions (adapted from Baxter et al., 2012)

Problem	Possible solutions
Negative perception of mask	Explore any underlying fears/preconceptions; offer a range of masks to try, allowing patient choice; hold mask in place initially so patient feels in control rather than using headgear; emphasise potential benefits of using the treatment
Concerns over operating machine and/or altering settings	Reassurance that machine settings are locked and cannot be altered accidentally; repeated practice under supervision operating machine; provide written/visual information to reinforce training
High-pressure tolerance noise/disturbance at night; Mouth dryness; leaks from mask	Use of ramp; use of rise time; ensuring no leak from mask. Place machine on towel to dampen any sound; check for leak from mask; ear plugs. Use full face mask; humidifier; drinks available; artificial saliva sprays/gel. Variety of masks to try to ensure best fit; replace cushion and/or headgear; check when the leak occurs, when put on or during sleep; excessive facial hair may need removing.
Problems securing clips and headgear	Explore alternative masks for ease of use; swap headgear from one mask type to another if easier to attach; consider using oral interfaces.
Soreness of bridge of nose or other areas due to pressure from mask	This can indicate that the mask is too tight or a poor fit and needs reviewing. A different interface/ type of face mask or strapping may be needed. Silicone gel can be helpful.
Problems wearing glasses or false teeth	Explore alternative mask designs which do not restrict the wearing of glasses. When fitting the mask check whether the user wears their teeth at night and fit the mask for the situation.
Restricted physical closeness	Reassure patient and partner that mask does not have to be worn all night, sometime off is okay.
Scratching an itch	Simply lift the mask off the face from the bottom if needed.
Excessive saliva or phlegm and poor cough strength	Maximise medications used to treat these problems; consider the use of lung volume recruitment bags, or a mechanical insufflator/exsufflator. Reassure patient and carer that sometimes these problems can make using NIV very difficult but some time may be better than none; consider daytime use when help is more available.
Concern about being unable to remove the interface because of arm and hand impairment	Use nasal interface Put in place emergency call system
Claustrophobia	Explore possible reasons for claustrophobia; use masks designed for claustrophobic people; use of mouth piece to familiarise patient with pressure sensation first; use of ramp; use of timed spells on the NIV (1 minute, 5 minutes, 10 minutes and so on)

## 2. Established assisted ventilation

A patient may initiate conversation about their future with many different professionals involved in their care. Similarly family members may do this also. Not all professionals will feel confident and have the necessary skills to discuss this. However, the minimum requirement for any professional caring for a patient with MND is that they have skills to pick up the 'cue', listen to concerns and thoughts and agree with the patient or family member how they would like to explore this further.

It is the role of senior nurses and doctors to explore understanding and wishes and to document these. However, others more involved in the day-to-day care of the patient can often instigate the involvement of senior professionals in this and the importance of this should not be undervalued.

More commonly it seems that patients expect professionals to initiate discussions about end-of-life care and wishes. Professionals, however, are often reluctant to bring this up, thinking it may upset patients, and wait for a lead from the patient. This can leave a crucial gap in patient care. Professionals have a responsibility to offer discussion about future wishes to patients.

Some phrases that could be used to initiate discussion about decision making and assisted ventilation:

- I wonder if you have been thinking at all about what might happen in the future?
- How are you finding your NIV?
- Have you ever thought about not using the NIV?
- What concerns you most, if anything, about your NIV in the future?

Deciding in advance what you would want to do in the future is hard and evidence would suggest patients often vary from their advance plan in their actions and decisions when the real situation arises.

One important benefit of advance care planning discussions is that it provides patients with the information they need to make a decision and allows them time to weigh this up before the situation arises. This allows the patient to make a more considered rather than spur-of-the-moment decision. So, although a patient may not know themselves in advance if or when they want to stop assisted ventilation, having had discussions in advance of this point they then may be more able to recognise this time and make a decision when that time arises. What seems like a sudden decision therefore can be based on months of mulling things over.

Advance care planning discussions may identify the things that are of great value or importance to the patient, such as wanting to be cared for at home; being able to say/control what happens to them. For patients with MND there are often levels of disability that they consider equate to an intolerable quality of life. Common examples with respect to deciding to stop assisted ventilation are 'when I can no longer communicate' or 'if I can do nothing for myself'.

Documentation about what has been discussed with a patient and their family and what their views and wishes are is of immense importance in supporting the care the patient wants, especially in times of crises and when professionals are called who have not met the patient before.

The discussion can help patients prepare for the decisions that they will actually take in the future. The record of the discussion helps professionals understand how the decision has been shaped and how confident and settled the view of the patient is. In the specific context of a patient who may wish to withdraw assisted ventilation at a time in the future, it also allows the team to begin to develop an appropriate plan of who, how and where and to involve people at an early stage to support this plan.

These discussions and future wishes with the patient will seldom be about the potential of withdrawal of assisted ventilation in isolation from other factors related to quality of life and end-of-life care. Discussion about what to do with the assisted ventilation should be a normal, integrated part of end-of-life care discussions and planning. This discussion may also usefully include resuscitation status and preferences for place of care.

The written record should include:

- date
- name of professional
- summary of content of discussion
- views of the patient about overarching values re: living (quality, spiritual duty, quantity), ceilings of care, interventions and care scenarios
- Thoughts about if and under what circumstances the patient would wish to stop assisted ventilation.

The patient may wish to construct an ADRT or appoint an attorney for decisions about life-sustaining treatment; direction and support should be offered for that.

These discussions should be revisited with the patient on a number of occasions and the record updated accordingly, affirming their continued view or documenting any change in wishes.

Discussions with patients about their advance wishes should be shared within the clinical team and decisions made available more widely as per local procedures in case of emergency. In Scotland, Electronic Key Information Summaries are enabling this communication and Electronic palliative care co-ordinating systems (EPaCCS) may aid this in England and Wales in the future.

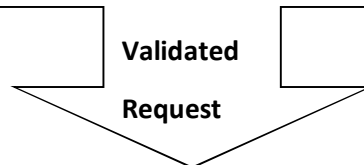
In almost all cases, the information should be kept with the patient. Most patients and families find this reassuring and helpful; only very occasionally would the patient or their family find this too distressing. Templates that may be used for emergency healthcare plans, statements of advance wishes and ADRT are available and signposted in Appendix 1.

#### Appendix 4. Specimen checklist for assisted ventilation withdrawal

Patient details / sticker	Responsible doctor:  Name:  Role:
---------------------------	---

##### One of the following criteria met

Patient with capacity has made a decision to stop assisted ventilation	<input type="checkbox"/>
OR	
Patient has a valid and applicable ADRT	<input type="checkbox"/>
OR	
Patient has appointed a LPAHW and a case of best interest is made	<input type="checkbox"/>



- |  |                          |
|--|--------------------------|
| Senior clinician appointed to co-ordinate withdrawal   | <input type="checkbox"/> |
| Discussion with patient and family where, when and how | <input type="checkbox"/> |
| Discussion with MDT why, where, when and how           | <input type="checkbox"/> |
| Contact home ventilation team if not already involved  | <input type="checkbox"/> |
| Care plan for removal of assisted ventilation agreed   | <input type="checkbox"/> |

##### At home

- |  |                          |
|--|--------------------------|
| Contact GP to arrange prescription of symptom-control medication | <input type="checkbox"/> |
| Equipment (see below)  | <input type="checkbox"/> |

##### After death

- |   |                          |
|---|--------------------------|
| Collection of equipment and drugs organised | <input type="checkbox"/> |
| Audit form completed                        | <input type="checkbox"/> |
| Notify key people about the death           | <input type="checkbox"/> |

##### Equipment list

Medications and water for injection; needles, cannulae, butterfly, syringes and syringe pump  
clinical waste bags for circuit & mask / tracheostomy tubes; sharps bin; dressing pack; scissors  
gloves.

## Appendix 5. What to do when a patient dies with NIV in use

It is not unusual for a patient to die whilst actually using their NIV. This can cause some anxiety for nursing and care staff and families. The patient's chest may still move with the backup rate of inspiration provided by the ventilator and the ventilator may alarm. The patient will not have a pulse or heart sounds and pupils will be fixed and dilated.

This should be explained to the family and to those who are providing care for the patient prior to the patient's death. Staff caring for a patient may need specific guidance as to what to do in these circumstances. An example care plan is given below.

This example care plan shows the NIPPV ventilator, which is only one of several models that patients may be provided with. A care plan devised for a specific patient will require indication of processes for the specific model of ventilator in use for that patient.

### *Example care plan for removal of NIV by care agency staff*

Patient details/sticker

#### **Introduction:**

This care protocol concerns the removal of the mask and cessation of non-invasive assisted ventilation (NIV) in the event of [Name]'s death.

#### **Purpose:**

In the event of [Name] dying, whilst the NIV is in situ, the ventilator will continue to work giving the appearance that [Name] is still breathing.

Support workers are not usually qualified or able to remove the NIV mask and turn off the ventilator.

A qualified professional must attend to verify that death has occurred and remove the NIV. **The ventilator should not be turned off and the mask should be left in situ until death verification.**

#### **Plan:**

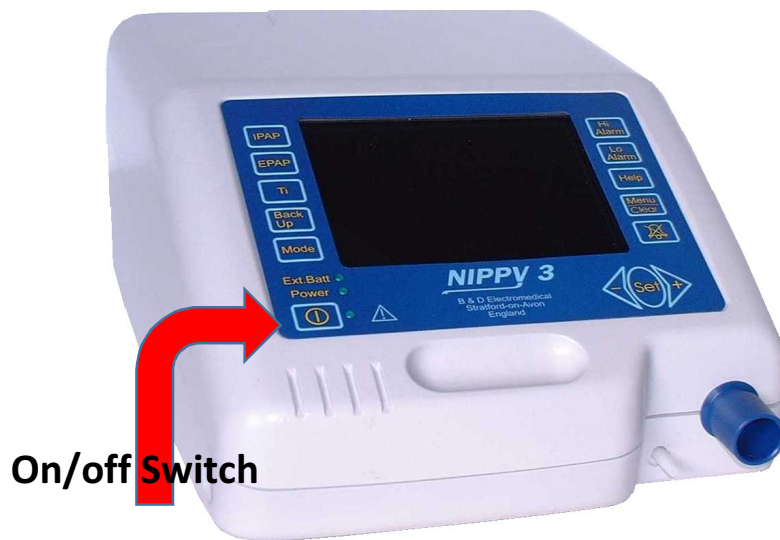
1) If the support workers suspect that [Name] may have died, ring the GP, Out of Hours Service or Ambulance Service. The decision around who to call will be determined by time of day and availability of the GP to attend.

2) The professional verifying death should turn off the ventilator and remove the mask as follows:

- Disconnect straps and remove face mask
- **Turn off** the ventilator by **pressing the on/off switch** which is at the bottom left hand side of the ventilator (Picture 1) and **hold it down until a red sign appears** in the centre of the screen.
- The red sign will ask if you want to turn the ventilator off.



- **Press and hold the on/off switch down** until ventilator stops.
- The mask and tubing can all be disposed of in general domestic waste. Contact hospital or designated provider to arrange for collection of the ventilator when appropriate.



## **Appendix 6. Audit of process and outcomes**

### **Background**

The purpose of this audit is to provide information that can lead to the improvement of care for patients and their families.

Whilst the Guidance is specifically for patients with MND, there is potential that the principles and specifics may be applicable to patients with other conditions causing respiratory failure and for this reason the data collection seeks to include any ventilator-dependent patient who requests that their assisted ventilation be stopped. It is hoped that this may inform guidance for other populations in the future.

This includes patients with:

- Motor Neurone Disease
- Chronic respiratory disease
- Duchene muscular dystrophy
- spinal injury
- other neuro-muscular and lung pathologies

The data will be analysed by a joint audit group including members of the Association for Palliative Medicine, British Thoracic Society, Home Ventilation UK, and Association of British Neurologists, and reported as anonymised information that can inform guidance and practice. Summarised, anonymised benchmarking data will be available to individuals and professional organisations. Non-attributable information may also be submitted for publication in peer-reviewed clinical journals.

### **Drug regime for the management of symptoms**

As part of developing guidance, we want to understand in as much detail as possible the drugs and the doses utilised in managing the symptoms related to withdrawing assisted ventilation. This will not be the same for each patient, but we need to understand the breadth of practice and how practice relates to outcomes.

We would be grateful if you would try to provide information for the following questions as best you can, with as many comments/provisos/qualifiers as you feel you need.

Your personal details will be used only to provide you with reports and benchmarking data. All reports will be anonymised and all publications non-attributable.

A Microsoft word and spread sheet version of the form is available on the APM <http://apmonline.org/publications/>

The completed audit form should be sent to [LAR.ventilation@nhs.net](mailto:LAR.ventilation@nhs.net)

Or by post or Fax to

Professor Christina Faull, Chair of the joint audit group,  
LOROS , Groby Road, Leicester LE3 9QE  
Fax 0116 231 8457

Your name:

Your job/role:

Your email:

### Section 1: Background Information about the patient

1.	Age of patient (tick one)	<30 30–50 50–70 >70
2.	Sex (tick one)	Male Female
3.	Diagnosis (tick one)	MND COPD DMD Cervical spinal cord injury Other (specify)
4.	Date of death	MM/YYYY
5.	What type of assisted ventilation was withdrawn? (Tick one.)	NIV (mask/non-invasive ventilation) IV (ventilation via tracheostomy)
6.	How long had the patient been on this type of assisted ventilation? (Tick one.)	>1 year 6 months–1 year 1–6 months <1 month
7.	Where did the withdrawal take place? (Tick one.)	Home Hospice Hospital (specify type of ward) Care Home
8.	Did the patient have capacity to make the withdrawal decision, or was this carried out as part of an ADRT (advance decision to refuse treatment) or 'best interests' decision?	Capacity ADRT Best interests decision
9.	Which doctor(s) had discussed and agreed with the patient and family the decision to withdraw assisted ventilation? (Tick all that apply.)	GP Cons Neuro Cons Pall Med Cons Resp/Home Vent Team Other (specify)

### Section 2. Information about the clinical picture in the day before assisted ventilation was withdrawn

10.	How many hours a day was ventilation in use (tick one)?	Overnight only <16 hours/day 16–22 hours/day >22 hours/day N/A
11.	How long could the patient manage without assisted ventilation support? (Tick one.)	Cannot manage at all A few minutes Up to an hour A few hours
12.	How did the patient communicate in their last days? (Tick one.)	Speech Eye movements Writing/keyboard They could not Other (specify)

13.	What was the patient's level of independence and function? (Tick one.)	Able to walk Mobile with use of wheelchair Bed- or chair-bound	
14.	Could the patient use their hands for any tasks? (Tick one.)	Yes No	
15.	What was the level of consciousness in the last days before withdrawal was commenced? (Tick one.)	Fully Alert Drowsy, responding to Voice Very drowsy, responding to touch/Pain Unresponsive N/A (locked in state)	
16.	In your assessment, what symptoms was the patient experiencing on the assisted ventilation in their last days? (Grade each 0–10.)	Breathlessness: Anxiety: Distress: Other (specify) :	
17.	What were the ventilator settings (prior to the withdrawal process)? (Fill as applicable.)	Mode of Ventilation  IPAP EPAP	Pressure control Pressure support Other  cm H2O cm H2O
18.	Was the patient already on an infusion (syringe driver) before the withdrawal of assisted ventilation was planned? (not started as part of the withdrawal plan. See Q21)	Yes No  If yes, specify details of drugs: Drug 1: Dose/24hr:  Drug 2: Dose/24hr:  Drug 3: Dose/24hr:	
19.	Before the withdrawal of assisted ventilation was planned, was the patient taking regular oral, transdermal or per gastrostomy opioid and/or benzodiazepine?	Yes No  If yes, specify details of drugs  Opioid: Dose/24hr  Benzodiazepine: Dose/24hr	
20.	Prior to the start of the withdrawal process (e.g. the night before the scheduled withdrawal) did you reduce the ventilator settings in anyway?	Yes No  If yes, please state in as much detail as possible what you did?	

21.	Prior to the start of the withdrawal process (e.g. the night before the scheduled withdrawal) did you increase drugs for symptom management in anyway?	Yes No  If yes, please state in as much detail as possible what you did?
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### Section 3. Information about the withdrawal

22.	What healthcare professionals were there to initiate the withdrawal (give professional role not names: e.g. GP, specialist ventilation nurse)?	
23.	Which healthcare professional took the lead in managing symptoms?	
24.	How long had the lead person known the patient for? (Tick one.)	Days Weeks Months Years
25.	Which healthcare professional specifically took the role of withdrawing the ventilator/taking the mask off? Or was this a family member?	
26.	What was the intention of symptom management before removing the assisted ventilation? (Tick one.)	To achieve total loss of awareness (sedation)  To make sleepy but still aware  No immediate symptom management was needed before withdrawing assisted ventilation  Other (specify)
27.	Did you give any medication (additional to any mentioned in Q18, Q19 or Q21 above) before you commenced withdrawal (i.e. anticipatory symptom management or sedation)?	First dose drug 1: Dose:  First dose drug 2: Dose:  First dose drug 3: Dose:  First dose drug 4: Dose:
28.	What route(s) for administration of drugs did you use? (Tick as applicable.)	IV SC IM PO

		Buccal Per-gastrostomy Rectal
29.	Was further medication needed to manage symptoms <b>before</b> the assisted ventilation could be fully withdrawn? (Fill in each as needed.)	Drug 1: Number of additional doses: Total Dose (including first dose in Q27):  Drug 2: Number of additional doses: Total Dose (including first dose in Q27):  Drug 3: Number of additional doses: Total Dose(including first dose in Q27):  Comments:
30.	How long before you withdrew assisted ventilation did you give the first dose of medication? (Add number of minutes/hours.)	Minutes Hours N/A
31.	How did you judge that symptoms were well enough managed to stop the assisted ventilation? (Tick one or add free text.)	The patient looked calm The patient was drowsy but awake The patient was asleep/lightly unconscious The patient did not respond to voice The patient did not respond to touch/pain The patient had lost corneal reflex Other
32.	Did you decrease the ventilator settings before completely stopping assisted ventilation?	Yes No  If yes, please state in as much detail as possible what you did?
33.	Was further medication administered to manage symptoms <b>after</b> the assisted ventilation was withdrawn? (Fill in separately for each time additional drug(s) were administered adding more similar records if required.)	1. Reason for further medication:  Drug(s) : Doses: Approximate time after assisted ventilation stopped:  2. Reason for further medication:  Drug(s) : Dose: Approximate time after assisted ventilation stopped:  3. Reason for further medication:

		Drug (s): Dose: Approximate time after assisted ventilation stopped:  Comments:
34.	Please summarise the drugs used to manage symptoms during withdrawal in Q27, Q29 & Q33.	Drug 1: Total Dose: Drug 2: Total Dose: Drug 3: Total Dose: Drug 4 : Total Dose:
35.	Were there any symptoms that were very challenging to manage effectively during withdrawal?	Yes No  If yes, specify and comment:
36.	Did the patient die with the mask/interface still in place?	Yes No
37.	Was the patient conscious after the assisted ventilation was withdrawn?	Yes No
38.	How long after the assisted ventilation was withdrawn did the patient live for? (Complete one.)	minutes hours days
39.	Were there any challenges related to family reactions during the withdrawal?	Yes No  If yes, please specify:
40.	What is your perception of what the experience was like for the family? (Tick one.)	Positive Difficult; beyond your expectation of normal grieving Frankly traumatic  Comments on issues/ how it could be improved:

#### Section 4. After the withdrawal

41.	Was there any immediate feedback from the family about the withdrawal if they were present, or anything they specifically commented on that may help others to know in the future?	Yes No  If yes, please specify:
42.	What was the experience like for you?	Positive Neutral Difficult Frankly traumatic

		Please comment on what made the process difficult or traumatic for you:
43.	Is there anything you would do differently next time, anything that could have gone better, or any learning outcomes to share?	Yes No  If yes, please specify:
44.	How has this affected your confidence in this area of care? (Tick as applicable.)	My confidence has increased My confidence is unchanged My confidence has reduced  I would prefer not to do it again
45.	Where there any issues that arose in the team debrief?	Yes No N/A no team debrief  If yes, then specify:
46.	Please add any other comments about the process of the withdrawal and symptom management	

**Thank you very much for taking part in this audit. Your contribution and time is very much appreciated.**





## Ventilation for motor neurone disease

If your breathing grows weaker with motor neurone disease (MND), your respiratory team may suggest using ventilation, where a machine helps support your breathing.

This information sheet explores the different types of ventilation and what to think about when deciding whether to use this support or not.

All quotes are from people living with or affected by MND.

### 1: How can ventilation help me?

If suitable for you, ventilation can help improve your quality of life by:

- relieving some of the symptoms caused by the weakening of your breathing muscles
- enabling you to breathe more effectively
- reducing fatigue
- reducing anxiety and distress.

However, everyone has a different experience with MND and the benefits may vary. It is also important to understand the different types of ventilation and how these will affect you in the future.

Ventilation may prolong life, but it will **not** prevent weakening of the breathing muscles or stop the progress of the disease.

### When do I need to think about ventilation?

Deciding whether or not to use ventilation can be a difficult choice. It is important to discuss the options with the respiratory professionals involved in your care, so that you can make an informed decision.

Your choice will depend on:

- your own views and preferences
- assessment to see if treatment is suitable for you
- the best timing for ventilation to be introduced
- your needs at that point
- what your wishes might be for your future care.

If possible, find out as much as you can about the options for ventilation as soon as you feel ready to do so.

*“This information sheet may be useful to show to your health and social care professionals, who may not always have experience of ventilation with MND.”*

It is helpful to have discussions with your health and social-care team before you develop any symptoms, or at the first sign of any changes to your breathing.

You may also find it helpful to discuss the options with your family and anyone involved in your care, as soon as you feel ready to do so.

This will give you time to:

- think about your choices, without a sense of urgency
- understand what your choices mean now and in the future
- ensure your wishes and preferences are known
- ensure everyone who supports you is prepared for the changes ahead, whether or not you decide to use ventilation
- avoid unwanted or unplanned interventions.

*“My own experience has shown that people are not always prepared early enough. Or symptoms have not been spotted early enough. In my husband’s case, his breathing problems were too far advanced for him to use non-invasive ventilation effectively.”*

## **2: What types of ventilation are available?**

There are two types of ventilation:

**Non-invasive ventilation (NIV):** where a machine supports your breathing by helping to boost your intake of normal air through a mask. This usually covers either your nose, or your nose and mouth, depending on the type of mask you find most comfortable.

The National Institute for Health and Clinical Excellence (NICE) has produced a guideline: CG105 on the use of non-invasive ventilation for people with MND. This document may help when discussing concerns about breathing with your respiratory team or wider health and social-care team. For details, see: Information Sheet 14C – *NICE clinical guideline on non-invasive ventilation (NIV)*.

**Invasive ventilation (also known as a tracheostomy):** where a tube is inserted into your windpipe through the front of your neck, which enables a ventilator to take over your breathing.

Using ventilation may not be suitable for everyone. If appropriate, it may help to relieve breathing problems, improve sleep and reduce fatigue, but it will not stop the progress of the disease.

### **What is full ventilation?**

Ventilation is usually needed overnight at first, but as the disease progresses you may need it for longer periods during the day. If you need to use ventilation for more than 12 hours in every 24, this usually means you are becoming dependent on the machine.

Either type of ventilation can be used part-time if you can still breathe when unsupported. However, invasive ventilation is often used on a continuous basis from the point of introduction.

If you need either type of ventilation continuously, it is called full ventilation. When you are fully ventilated, you are likely to become reliant on this support. Without it, you will become very breathless in a short time and may be unable to breathe effectively on your own.

### **What happens if I decide not to use ventilation?**

If ventilation is not suitable for you or you decide not to use it, your respiratory team, physiotherapist and other professionals, such as your palliative care team, can advise on other ways to help. This may include:

- posture and positioning
- breathing exercises
- assistance if you find it difficult to cough
- ways to relax
- medication to ease symptoms and anxiety.

For details about other breathing therapies, see: Information sheet 14A – *Understanding how motor neurone disease might affect breathing*.

### **Do I need extra oxygen?**

In most cases, ventilation for MND uses normal air to help you breathe. Extra oxygen is not usually recommended with MND, as it can upset the balance in your body between oxygen and carbon dioxide.

However, if your oxygen levels are low, it may sometimes be used with caution. Any decisions regarding oxygen for home use should be discussed with your respiratory team, as high levels of oxygen in your blood can be harmful.

For details about the use of oxygen with MND during air travel, see: Information sheet 14E – *Air travel and ventilation for motor neurone disease*.

## **3: How does non-invasive ventilation work?**

NIV boosts the flow of normal air into your lungs through a mask that covers your nose, or nose and mouth. This is attached by tubing to a small machine, powered either from a normal electric socket or a battery.

The air flow from the NIV machine is timed to match your normal breathing pattern. Some machines adjust the timing automatically.

### **Where can I use NIV?**

There are several different types of machine, but they are small, portable and can be used anywhere, including at home and on the move.

Even if you need to use a wheelchair, you can still be mobile, as some of the machines can be powered by a battery. If travelling by car, some machines can be plugged into the cigarette lighter.

For details about taking NIV onto an aeroplane, see: Information sheet 14E – *Air travel and ventilation for motor neurone disease*.

The respiratory team will show you how to use the machine. The settings, masks and other parts of the ventilator will be adjusted to suit you. This is usually carried out at hospital as an outpatient, but may require a short stay. However, some of these services may be able to see you at home.

Your usage of the ventilator will be regularly reviewed in case your needs change. The team can provide ongoing support, including advice about maintaining and cleaning the equipment, and out-of-hours urgent support if needed. They may be able to arrange visits at your home if you find it difficult to travel.

Getting used to your machine and the mask does take time. You may adapt very quickly or you may need to try different settings or masks. Your respiratory team will provide support, but if you decide it is not right for you, they may be able to offer other options to help manage your symptoms.

### **When is NIV not suitable?**

Your assessment with the respiratory team is important, as there are various things to consider.

NIV may not be suitable if:

- you do not have either a paid or unpaid carer for long periods during the day or night and you have weakness in your arms or hands – this means you could find it difficult to put on or take off the mask
- you feel claustrophobic or very sore when wearing the mask
- you cannot adjust to the way the flow of air feels when using the machine
- you have swallowing difficulties.

### **What happens over time with NIV?**

At first, you may only need to use NIV at night, to help improve your breathing and quality of sleep. However, as the disease progresses, you may need to use it during the day to help relieve your symptoms.

*“I’m at the point where I am using NIV for 15 hours a day.”*

This means you can become reliant on NIV over time and may reach a point where you need to use it constantly. However, it is not the same as life support.

Even with NIV, there will come a time when the breathing muscles become too weak for the machine to help. For some this can take many months, for others it may happen more quickly. This leads to drowsiness, unconsciousness and ultimately death. This is usually peaceful and your health and social-care team can support you to reduce anxiety (see later Section, 5: *Can I stop using ventilation?*).

In this situation, resuscitation is unlikely to be helpful, as your NIV will not be able to support you effectively.

Invasive ventilation works in a different way, as it breathes for you (see next Section, 4: *How does invasive ventilation work?*)

## **4: How does invasive ventilation work?**

Invasive ventilation uses a tube inserted through the front of your neck into your windpipe to help you breathe and enable suctioning of secretions. The insertion of the tube is called a tracheostomy, which is provided under general anaesthetic. The ventilation machine uses this tube to help you breathe.

Invasive ventilation is not always available or offered to people with MND in the UK, but it is sometimes used if NIV is not suitable.

This type of ventilation usually means being fully ventilated and care can be complex. This requires support from an experienced multidisciplinary team and those involved in your care.

### **What do I need to think about with invasive ventilation?**

A facial mask is not required with invasive ventilation, which means you can avoid claustrophobia or discomfort. However, there are other things to consider.

Like NIV, it is portable and can be used at home, but your family and carers may need to provide increasing levels of support. This can be challenging for them and frustrating for you, which could continue for some years with this type of ventilation. Invasive ventilation is most successful where your other symptoms of MND are progressing slowly and your quality of life is good.

If the level of care you require increases, it may no longer be possible to support you at home and may mean moving into a nursing home. In certain circumstances, invasive ventilation may only be offered if you are prepared to move into a nursing home, due to the complexity of care.

If MND causes problems with your speech and communication, invasive ventilation can make this more difficult. Your speaking pattern can be disturbed by the rhythm of the ventilator and with some machines it may be difficult to speak at all.

However, speech and communication problems with MND will continue to progress with or without ventilation, so it may become more difficult to tell people about your wishes for future care. It is important to plan ahead to ensure everyone involved in your care is aware of your wishes (see later Section, 6: *How do I plan ahead for my future care?*).

Ask your respiratory team for advice on all these aspects. With invasive ventilation, you may also want to ask about:

- maintenance of the equipment, as this requires input from professionals
- replacing the tube, which is usually done once a month (this is a minor procedure, but does carry some risk)
- suctioning mucus from the airway, which is needed more frequently in the first few weeks of ventilation and whenever chest infections occur
- daily or more frequent care to prevent blockages, infections and skin breakdown around the tubing.

Whether or not to use any type of breathing support is your decision. However, in unexpected situations, it is possible for invasive ventilation to be introduced without your prior consent.

For example, if you are resuscitated in an emergency, it may be used to help you recover. This is usually temporary, but the emergency team may not realise how difficult it can be to stop using invasive ventilation once breathing muscles have weakened with MND. You may then have to accept this type of ventilation as ongoing support, which can affect plans for your future care.

If invasive ventilation is something you definitely do **not** want in any circumstances, you need to make this clear to all those involved in your care. You may need to write down your wishes to guide people if you become unable to make decisions or communicate for any reason (see Section 6: *How do I plan ahead for my future care?*).

## **5: Can I stop using ventilation?**

You can stop using ventilation at any time. It is your legal right to ask that a treatment like breathing support be stopped.

You may wish to stop using ventilation if you feel it is no longer helping or has become a burden. If you use it only some of the time, you may choose not to put the machine back on after a gap. You will probably need other therapies to manage your symptoms, so it is usually best to plan how you will stop with your health and social-care team.

However, coming off ventilation is very difficult if you already need continuous support. If you are fully ventilated and can no longer breathe effectively on your own, a natural death is likely to follow in a fairly short period of time. Your decision to stop must be made with the clear understanding that it will cause a significant risk to your life. Discuss this with your respiratory team or palliative care professionals, who will explain how medication can help you feel calm and relieve distress if you decide to stop in these circumstances.

Your respiratory team, palliative care team and wider health and social-care team can answer any questions you may have about planning ahead, including how to manage symptoms and support for your family.

You can also record advance decisions to stop using ventilation in specific circumstances, in case you become unable to choose or communicate for yourself (see Section 6: *How do I plan ahead for my future care?*)

## 6: How do I plan ahead for my future care?

Using breathing support can raise questions and fears about the way MND will progress. This may be a good time to open conversations with your family and health and social care team about your future care. This can reduce anxiety and you may find it helpful to talk about:

- choices for end-of-life care
- options for withdrawal of ventilation, if it is no longer helping or has become a burden
- what will happen in the later stages of MND, as knowing the facts can help reduce fear
- how to record your wishes about future care.

Sharing your thoughts and decisions with everyone involved in your care helps them to meet your needs and wishes. It can also help put your mind at rest.

It is important to have these conversations as early as you can. Speech and communication can be affected by MND and some people also experience changes to thinking and reasoning. This means it may become more difficult to have complex discussions.

In case you become unable to make decisions or communicate, you can record your wishes about future care and treatment. This helps others to understand how you want to be supported and anything you do not want to happen.

This is usually done using one or both of the following:

- **Advance care plan:** this enables you to record your wishes about any aspect of your future care, treatment or practical assistance you might need. It is not a legally binding document, but helps guide everyone involved in your care.
- **Advance Decision to Refuse Treatment (ADRT):** this enables you to record which treatments you do not want introduced or any that you want to be withdrawn under specific circumstances in the future. For example, you may wish to have NIV withdrawn at a particular point. If completed correctly, and you can show that you are able to make reasoned decisions when it is created, your ADRT is legally binding.

As your symptoms progress, your wishes may change. You can review and amend your advance plans or decisions at any time. For details about how to plan ahead and make advance decisions, see *Further information* at the end of this sheet about our end-of-life and ADRT publications

Our MND Connect helpline can provide a listening ear or guidance about future planning (see *Further information* at the end of this sheet for contact details).

The helpline team can also direct you to our Association visitors, branches, groups and regional care development advisers, who can all listen to your concerns and help you to find further information.

## **7: How do I find out more?**

We provide other information sheets related to breathing support and MND:

- 8A – Support for breathing problems
- 8C – NICE guideline for non-invasive ventilation
- 8D – Troubleshooting for non-invasive ventilation
- 8E – Air Travel and ventilation for motor neurone disease
- 14A – Advance Decision to Refuse Treatment (ADRT) explained

We also provide the following guides:

- *Living with motor neurone disease* – our main guide about MND and how to manage its impact
- *Caring and MND: support for you* – comprehensive information for unpaid or family carers, who support someone living with MND
- *Caring and MND: quick guide* – the summary version of our information for carers
- *End of Life: a guide for people with motor neurone disease* – our comprehensive guide to making decisions about future care and late-stage MND, including advance care planning and advance decisions

You can download most of our publications from our website at:

[www.mndassociation.org/publications](http://www.mndassociation.org/publications) or order in print from the MND Connect team, who can provide additional information and support:

### **MND Connect**

MND Association, PO Box 246, Northampton NN1 2PR

Telephone: 08457 626262

Fax: (01604) 638289

Email: [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

## Appendix 8. Glossary of Terms and Abbreviations

**Advance care planning (ACP)** is a process of discussing and planning ahead between a person and their care providers regarding the person's care in the future and at the end of life. Whilst it may result in documents that are useful and legally important if the patient loses capacity, significant benefits of advance care planning result from the sharing of information between professionals, patients and families enabling patients to be better placed to make decisions as they deteriorate without loss of capacity.

**Advance decision to refuse treatment (ADRT)** is the decision of a patient to refuse specific treatments offered to them relating to specific circumstances. When it is valid, it is legally binding.

**Advance directive** is now replaced by advance decision to refuse treatment. It is sometimes called a living will.

**Advance statement** is a general statement of views and wishes and allows the person completing the statement to indicate their preferences for receiving or refusing forms of treatment in the future. They may express these preferences in the form of a 'values history'. These documents are not considered legally binding, although they provide an opportunity for the person to express their wishes regarding their future care, which should be taken into account if best-interests decisions require to be made on their behalf in the future, should they become incapacitated.

**Anticipatory prescribing** in palliative care is the provision of medications for distressing symptoms prescribed and made available prior to the symptoms occurring. In the community setting this will mean that these medications are in the patient's house prior to their requirement.

**Family** in this guidance the use of the word 'family' is inclusive of those close to the patient as well as those related.

**IPAP** is inspiratory positive airways pressure.

**Lasting power of attorney (LPA)** is a legal tool for a patient in England or Wales to appoint someone to make decisions on their behalf if they lose capacity. In Scotland, the equivalent term is **welfare power attorney**. The LPA may be for health and welfare and/or for property and financial affairs. The LPA for health and welfare must be specifically given authority by the patient to make decisions about life-sustaining treatments.

**Locked-in state** is complete paralysis of voluntary muscles in all parts of the body except for those that control eye movement.

**Motor neurone disease (MND)** is a progressive neurodegenerative disease that attacks motor neurons leading to weakness and wasting of muscles, causing reduced power in the limbs and difficulties with speech, swallowing and breathing. Eye movements, sight, hearing, bladder, bowel and sexual function are unaffected. Intellect is preserved but some changes in cognitive function are common.

**Non-invasive assisted ventilation (NIV)** is a form of mechanical assistance with breathing that does not require the patient to be intubated. Air is given under pressure to the patient through a full face nasal mask, or mouth piece. Some patients use a combination of these interfaces.

**Respiratory failure** is inadequate gas exchange by the respiratory system resulting in either low oxygen levels, high carbon dioxide levels or a combination of both.

**Tracheostomy assisted ventilation (TV)** is mechanical assistance with breathing using a tube placed in the trachea, usually through a stoma (hole) in the neck, connected to a ventilator.



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