

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

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Association for
Palliative Medicine
Of Great Britain and Ireland

Endorsed by:

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Consultation

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Consultation

Guidance Summary and Standards for Care

The purpose of this Guidance is to support professionals in working with the small number of patients with MND who are dependent on their ventilator and ask that this assisted ventilation be withdrawn.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 the dying process, despite increasing dependence on its use.

However, a minority of patients with MND who are ventilator-dependent request that the assisted ventilation is withdrawn. 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.

Ventilator dependence can be defined as the use of assisted ventilation for more than 16 hours a day. There are degrees of ventilator dependence within this group of patients, some patients will be unable to tolerate even a few minutes without assisted ventilation and others will be able to tolerate several hours. If there is uncertainty regarding the degree of ventilator-dependence for an individual patient, then the following guidance can be applied.

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.
- It is a patient's legal and ethical 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 a clinical leader is required.
- 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 generalizable but the precise methodology requires individual tailoring to the patient.
- As this is a rare area of care with very little published evidence there is a need for ongoing evaluation of methods and outcomes.

This is an area of care that requires considerable preparation and planning which should be facilitated by a clinical co-coordinator. 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.

The process of care does not stop immediately after the patient has died. The family may require considerable ongoing support, especially opportunity to discuss events with the professionals who have been directly involved. The professional team may also require debriefing to acknowledge the impact of this uniquely challenging process. No one should be left with any doubts that the withdrawal of assisted ventilation at the request of a patient is both ethical and legal.

A Summary of the Guidance

Timing	Standard	Process to address standard (s)
When commencing assisted ventilation	<p>Standard 1 Patients should be made aware that they have the right to ask to stop assisted ventilation. They should be in no doubt that this is legal and that healthcare teams will support them.</p>	<p>Offer patients and families the opportunity to discuss future scenarios when assisted ventilation is being considered.</p> <p>Inform patients that they can stop the treatment at any time, that it is entirely their right and legal and that their health care team will manage their symptoms in a different way.</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>
Withdrawal of assisted ventilation	<p>Standard 2 A senior clinician should lead the planning and co-ordination of the withdrawal.</p> <p>Standard 3 Withdrawal should be undertaken within a few days of an affirmed request.</p> <p>Standard 4 Symptoms of breathlessness and distress should be anticipated and effectively managed</p>	<p>Discuss with the patient and family when, where and how this will happen, including the potential for living for some hours without the ventilator.</p> <p>Discuss with the professionals when where and how this will happen, identify key people and their roles. Ensure members of the MDT understand the ethical principles and the legal position.</p> <p>Affirm the decision by assessing the patient's capacity or validity and applicability of an ADRT and that this is a settled view; allowing a period of time for discussion and reflection between the initial and the final patient's decision.</p> <p>Make a plan for symptom management: - Does the patient require sedation (ventilator dependent patients) before assisted ventilation withdrawal or</p>

		<p>augmented symptom control (patient can manage sometime off assisted ventilation)?</p> <ul style="list-style-type: none"> - What drugs, doses, route? - Who will prescribe and administer? - Who will manage the ventilator and how will settings be adjusted and mask/tubing removed? <p>Administer anticipatory medication, titrating opioids and benzodiazepine to manage symptoms.</p> <p>For those who are ventilator dependent assess effectiveness of symptom management by reducing assisted ventilation for a short time before full removal.</p> <p>Continue to titrate opioids and benzodiazepine to manage symptoms.</p>
<p>After death</p>	<p>Standard 5 After the patient’s death family members should have appropriate support and opportunities to discuss the events with the professionals involved.</p>	<p>Consider the needs of family and professionals after death:</p> <ul style="list-style-type: none"> -Plan who will provide support to family. - Debrief for professionals/ significant event analysis. <p>Submit data set and share key learning.</p>

A list has been compiled of senior health care 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 Association for Palliative Medicine (becki@compleat-online.co.uk)[others to be added] and it is available to anyone who feels this would be useful.

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. A high percentage of 76 palliative medicine doctors who had experience in withdrawing assisted ventilation rated the practical, ethical and emotional challenges as 7 or more on a 0-10 scale. Most doctors had only undertaken this on a single occasion. Lack of guidance on practical aspects of withdrawal, poor

advance care planning, lack of experience and the need to support all involved 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 the patient and they 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 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 smaller number of patients who use continuous tracheostomy delivered mechanical 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 other reason for loss of capacity.

The purpose of this Guidance is to support professionals in working with ventilator-dependent patients with MND who ask that their assisted ventilation be withdrawn and in the anticipatory management of their acute and severe dyspnoea.

The aim 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. Through ongoing collection of a minimum dataset described in this Guidance 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.

The Guidance is specifically for patients with MND. However there is potential that the principles and specifics may be applicable to patients with other conditions who request withdrawal of assisted ventilation or for whom assisted ventilation is no longer clinically appropriate. For this reason the audit (Appendix 6) intentionally goes beyond collection and collation of data from patients with MND to include any ventilator-dependent patient who request that their assisted ventilation be stopped. It is hoped that this may inform guidance for other populations in the future.

The Guidance is aimed primarily at professionals but we recognize that some of the content may also be useful to patients, families and others in supporting people with MND.

Association for Palliative Medicine Position Statement April 2015

Withdrawal Of Ventilatory Support At The Request Of An Adult Patient With Neurological Or Neuro-Muscular Disease

This statement intends to set-out the legal and ethical position for the care of patients with neurological or neuro-muscular conditions in the UK who request that their ventilatory support be withdrawn. Whilst the ethical principles are generic and applicable across the UK, the law in relation to mental capacity differs between England and Wales combined, and Scotland. The position statement is not a clinical guideline.

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 may become a medically manipulated variable 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 registration of the death from the perspective of either coroner or the law (except in circumstances where the disease itself requires different).
6. Withdrawing a medical treatment that a patient with capacity no longer wants, even if this is considered life-sustaining, is not assisted suicide.
7. Withdrawing a medical treatment from a patient who no longer has capacity, but who whilst having capacity made an ADRT which is specific in this regard and valid for these particular circumstances, is not euthanasia even if the medical treatment is life-sustaining.
8. 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 (LPAPW), including decisions on life-sustaining medical treatment, and where on multidisciplinary review this request meets 'best interests' criteria, it is not euthanasia even if the medical treatment is life-sustaining.
9. Withdrawing a medical treatment from a patient who no longer has capacity but who has not made an ADRT or appointed an LPAPW, is a conventional 'best interests' determination;

the principles of which are set out within the MCA 2005 and refined within more recent case law.

10. 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 and the multidisciplinary team preferably should begin before assisted ventilation starts and continue throughout the duration of the illness.
11. 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.
12. 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.
13. 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.
14. 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.
15. 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.
16. 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: An overview of assisted ventilation and withdrawal in MND

MND is a fatal neurodegenerative disease. Patients vary in the way MND first affects them and in 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, particularly those with limb-onset disease. Guidance from the National Institute of Clinical Excellence (NICE) supports its use. Undoubtedly NIV has many benefits for patients and the majority of patients choose to at least try NIV. A few 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. At this stage, it is of vital importance that the patient starting 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 for control of sleep disordered breathing. 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 24hrs/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, despite increasing dependence on its use. 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 whilst still using it.

However a very small but potentially increasing number of patients (perhaps 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 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.

Some may make a written statement or directive with respect to withdrawal in advance of their losing the ability to communicate or other reason for loss of capacity.

Evidence suggests that too few patients know about their potential choices and are not asked about their views of continuing assisted ventilation. There is a clear need for more information sharing and improvement in facilitated decision-making.

Challenges in the withdrawal of assisted ventilation

Interviews with 67 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.
- Whilst 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 individualized 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 health care 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.

This work has resulted in recommendations for practice and care on which this Guidance is based.

The evidence base for symptom management during withdrawal of assisted ventilation

For patients who use TV or NIV most of the time the removal of the assisted ventilation is likely to result in a very rapid onset of distressing symptoms and anticipatory medication is needed to manage the physical distress. For those patients that can manage a few hours without assisted ventilation, responsive augmentation of symptom management will be needed.

There is a small volume of literature discussing the clinical and practical aspects of withdrawal of assisted ventilation in MND and some 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 evidence that opioids and benzodiazepines, in doses titrated to manage symptoms and distress, do not shorten life (and may paradoxically delay it) 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 of continuing to be alive 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 indicates 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. GMC Guidance 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

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 health care professionals, it may also have a direct effect on their practice. To achieve best outcomes for patients and their families the impact on professionals and the support they require to avoid adverse effects on their practice, needs to be anticipated and planned for.

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. We only have the documented views of the family members and professionals who have reflected on this.

Whilst 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 17 interviews with family members and with 50 health professionals 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 are often given little information about the withdrawal process, how long it might take for the patient to die and any symptoms they may expect to see (e.g. gasping, colour change). This can cause additional stress.
- Families are often not made aware of potential patient behaviours which may occur during withdrawal such as colour change and agonal breathing, and were sometimes shocked at methods of symptom control if these has not been discussed beforehand.
- Families are frequently asked or even expected to take a role in the actual withdrawal of the ventilator, and sometimes left alone during the process. This is a poor outcome and a plan for the professional roles in all aspects of care needs to be clear.
- 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 the health care professionals involved.
- Family members can feel very isolated and unable to discuss the situation before or after with their usual social support.
- Relatives may need additional support after the withdrawal and still have questions and issues they need to discuss with the health care 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 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.

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

Standard 1

Patients should be made aware that they have the right to ask to stop assisted ventilation. They should be in no doubt that this is legal and that healthcare teams will support them

The process of consent for assisted ventilation support options should include reassurance that assisted ventilation can be stopped at the patient's request whilst they have capacity, or in accordance with an Advance Decision to Refuse Treatment (ADRT) or the direction of a Lasting Power of Attorney for Health and Welfare (LPAHW) in England and Wales, or Welfare Power of Attorney in Scotland, in the event of losing capacity. There is a case for more formalised consent for initiation of assisted ventilation.

It is important to promote the concept of advance care planning with patients who use assisted ventilation, especially those who have lost one modality of communication (speech or writing). Even if this does not contain 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, where neither an ADRT nor LPAHW has been generated.

Patients should be reassured that if there comes a point for any of the above reasons 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 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).

Patients and their next-of-kin should also be reassured that this process is lawful and will not create any difficulty with issuing a medical certificate as to the cause of death (which is MND).

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, and Scotland or Northern Ireland. More extensive discussion of the ethical and legal principles and their application is included in Appendix 3.

Standard 2

A senior clinician should lead the planning and co-ordination of the withdrawal

The journey towards the decision to withdraw assisted ventilation is discussed in detail in Appendix 4. For some patients this potential decision has been known about for some time, for others it appears more sudden.

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. For a patient who makes an 'unanticipated' (to the professional team at least) decision, the MDT involved in their care needs to decide who will affirm the decision and co-ordinate the process.

The coordinator may be the:

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

Standard 3

Withdrawal should be undertaken within a few days of an affirmed request

For all patients the decision needs to be affirmed, ensuring that it is a settled decision of a patient with capacity or that the advance decision is valid and applicable. A period of time for discussion support and reflection with the patient and family will be needed to be sure that this is the patient's settled choice.

In all circumstances the family must also have an opportunity to share information, ask questions and express concerns. There should be discussion with the patient and family on at least two separate occasions and ideally involving two different senior health care professionals.

When a patient decides that they no longer wish to have assisted ventilation then the support for its withdrawal must be arranged as quickly as possible, but this cannot be immediate. When a patient has reached this momentous decision it is understandably distressing to both them and their family if actions are not taken quickly. Practically, in most part because of the need for professional availability for a period of some hours, there will be a gap between the affirmed request and the withdrawal. Patients and families need to be supported in this time. It would seem reasonable that this delay is in the order of a 3-7 days at most.

Evidence for the decision

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
- Who was involved in discussions
- Alternative approaches are known and rejected by the patient
- That the patient knows they will die as a consequence
- That there is no coercion or that the decision is driven by mistaken kindness to the family
- That this a settled view of the patient
- Capacity assessment
- Summary of the benefits and burdens (if applicable)
- Statement of best interests (if applicable).

See Appendix 3 for additional guidance.

1. What to plan for

A specimen care plan for this is provided in Appendix 5.

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?
- Who will confirm the patient has died to the family and the professional team members?

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 mean 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 48hrs (nurses)
- Coordination of professionals availability and family support (especially if family travelling to be there)

What will be done in practical terms? (See Section 4 below)

- What drugs will be used?
- How will drugs be delivered?
- What other equipment will required and how will it be organised?

- How will mask/tube and ventilator be managed?
- What will be done with tube/mask/equipment immediately after death?

2. What to discuss with patient and family

The elements of the discussion with the patient should also be had with the family, ideally at the same time but this may not always be desired by the parties concerned.

The difficulty and impact of the decision should be acknowledged and the ethical and legal position underpinning the withdrawal of assisted ventilation reaffirmed. The ethical and legal 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 what medications will be given and how they will be given
- Mechanics of withdrawal including who will do what
- What will happen once the mask has been removed? Acknowledge the uncertainty about rate of deterioration to death on stopping assisted ventilation. Allay expectations that this will be immediate and could be some hours
- Address fears about distress and what would happen should distress be evident
- Discuss what additional professional support will 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. The availability of a nearby room for family is important as often they need some space for 'time out'.

Address any other concerns or issues.

Is there anything else that is important to the patient at this time?

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 that occur when someone is dying, appearance, colour changes (some patients become very blue).

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

3. What to discuss with other professionals

A list has been compiled of senior health care 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) [others to be added] and it is available to anyone who feels this might be useful.

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 coordinator 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

4. Symptom management

Standard 4

Symptoms of breathlessness and distress should be anticipated and effectively managed

Withdrawal of assisted ventilation will lead to predictable breathlessness and distress, which should therefore be anticipated and managed proactively. For those patients who can tolerate only minutes without assisted ventilation these symptoms come on very rapidly and, without medication, may be severe from the start. For those that can tolerate longer periods without assisted ventilation these symptoms may increase immediately but for some patients, symptoms might increase more slowly in their severity.

Although each case will vary there are some common principles to consider.

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 difficulty of venous cannulation.

These principles will be considered in detail below. Since it is recognized there are a number of potential approaches to symptom management, case examples are given in Section 5 to demonstrate how these principles may be reflected in practice.

Level of ventilator dependence

Best outcomes result from consideration of patients into two groups:

Group S: Sedation

Those 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 minute ventilation for a short time and adjustment of medication as necessary. This can be done by reducing the inspiratory pressure (IPAP) by around 50%. For example from 15cm H₂O to 8cm H₂O. This can be done in more gradual steps if that aids titration of medication. The back-up rate may also be reduced.

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. A deeper level of sedation may be required in some patients in order to manage breathlessness or distress. Absence of eyelash response whereby gentle stimulation of the eyelash elicits no reflex in the lashes may indicate the level at which proportional but adequate sedation is achieved in this group.

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

Group ASC: Augmented Symptom Control

Those who can tolerate longer periods of time without assisted ventilation will develop symptoms after a longer period of time and therefore do not require sedation but 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.

In either group breathlessness may be less sitting up and become acutely severe on lying down. The position and place (bed or chair) that the patient is to be cared for needs to be thought about as part of the plan of care and symptom management.

Medication

Route of administration

Medication may be delivered by any route but intravenous (IV) administration arguably 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. However for the purpose of withdrawal of assisted ventilation this route may not give the desired speed and reliability of response.

Buccal and nasal administration are 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.

Generally the route chosen should be acceptable to the patient and one in which the team feel confident.

Agents

Opioid and benzodiazepine medication should be used to manage breathlessness and distress respectively and are 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 of 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 2nd 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 doses to achieve this. The appropriate dose may vary with age and physiological resilience.

To achieve sedation for opioid naïve patients who are ventilator dependent (Group S):

- SC: Morphine or Diamorphine 15mg with Midazolam 15mg stat as initial dose, assessed for effect 15 -20 minutes later. Further aliquots of 5mg to 10mg of both opioid and benzodiazepine titrated to level of sedation.
- IV: Morphine or Diamorphine 10mg with Midazolam 10mg stat as initial stat dose. Further aliquots of 2mg to 5mg each of both opioid and benzodiazepine every two minutes 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% and the back-up rate, or by temporarily switching the ventilator off for a few minutes, with adjustment of medication if necessary.

Further aliquots of 5mg IV or 10mg SC of both opioid and benzodiazepine should be given as required by symptoms.

To achieve augmented symptom control for opioid naïve patients who are less ventilator dependent (Group ASC):

- Morphine or Diamorphine 5mg to 10mg SC or 2mg to 5mg IV with Midazolam 5mg to 10mg SC or 2mg to 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 and the route of administration of the drug. If this is administered IV then giving it at time of mask removal will allow sufficient speed of titration of medication to any symptoms as they arise. If medication is used SC then the initial dose should be given 30 minutes in advance of the mask removal. Subsequent doses of SC medication will take at least 10 minutes to have effect so very vigilant monitoring for early signs of distress is crucial as there is a risk of under treating.

For a patient who is already receiving opioids the dose of drugs administered should be adjusted to the 4 hour equivalent plus a 50% increase.

Oxygen¹

When ventilation is stopped, hypoxia may develop rapidly and may contribute to increasing breathlessness and distress. In addition to medication approaches, oxygen (if available and acceptable to the patient and family) may offer some control of these symptoms and can be applied via a nasal cannula or via tracheostomy. The use of medication and oxygen may have a synergistic effect upon symptom control, but there is no evidence (other than anecdotal experience) to clarify this and assessment on an individual level is recommended.

¹ 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.

Assisted ventilation

Familiarisation with the ventilator is crucial before assisted ventilation withdrawal takes place.

It is vital to know how to:

- turn off or adjust alarm settings
- turn off the machine
- reduce pressure settings and back-up rate.

5. Case examples

These are anonymised real examples to help give an in depth insight in to what undertaking this care may look like and some view of variability in patient need and clinical approach.

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/24hrs was commenced the night prior to the withdrawal.

Time Zero: ZA took off his NIV and was given morphine 5mg + midazolam 5mg
30 minutes: Not sleepy enough: morphine 5mg + midazolam 5mg
50 minutes: Signs of mild distress. Not sleepy: morphine 5mg + midazolam 5mg
75 minutes: Sleepy but some signs of distress: 6.25mg levomepromazine
80 minutes: Died

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 2 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 lead to some distress. Midazolam 5mg
75minutes: 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 unexpectedly decided to stop her assisted ventilation one Sunday during a respite stay at the hospice. Three family members were with her together with a doctor and three nursing staff. The on-call doctor had known the patient for one day and was supported by the Consultant by phone.

She was on a SC infusion of oxycodone 50mg + midazolam 70mg/24hrs 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 + Levomeproamzine 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 her stopping her assisted ventilation. She was mildly breathless with NIV in place and had a SC infusion of midazolam 10mg with Morphine 5mg/24hrs.

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 H₂O. 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 pulse rate, increase respiratory rate)

100 minutes: Died

6. After withdrawal

Documentation

All the professionals involved should make appropriate documentation of:

- The decision-making process (see Section 1)
- A summary of the medication and other strategies for symptom management
- Who did what
- Patient related outcomes
- Family related outcomes
- Time of death.

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.
- 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 coordinator 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

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). The details for the audit submission process have yet to be finalized.

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 he/she 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.

References

The relevant law

Re B (Adult, refusal of medical treatment) [2002] EWHC 429 (Fam) 2 All ER449, Right of a patient who has capacity to refuse life-prolonging treatment.

Mental Capacity Act 2005: <http://www.legislation.gov.uk/ukpga/2005/9/contents> (England and Wales)

Re C (Adult refusal of treatment) [1994] 1 All ER 819

R v Bodkin Adams [1957] CLR 365 (Duty to relieve pain; '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')

House of Lords debate re Annie Lindsell *Hansard* HL 721-724 (Nov 20 1997) (Duty to relieve suffering and distress at the end of life with particular reference to MND)
http://www.publications.parliament.uk/pa/ld199798/ldhansrd/vo971120/text/71120-18.htm#71120-18_head0

Adults with Incapacity (Scotland) Act 2000: <http://www.legislation.gov.uk/asp/2000/4/contents>

Proposed new mental capacity legislation (Northern Ireland)
<http://www.dojni.gov.uk/Consultation-on-proposals-for-new-Mental-Capacity-Legislation-for-Northern-Ireland>

Guidance from the BMA, Department of Health and GMC

BMA: Withholding and Withdrawing Life-prolonging Medical treatment: Guidance for decision making. 3rd edition 2007. Blackwell Publishing.

Department of Health. Reference Guide to consent for examination or treatment. 2nd edition 2009
https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/138296/dh_10365_3__1_.pdf

GMC Consent guidance: patients and doctors making decisions together. 2008 Legal Annex- Common Law.
http://www.gmc-uk.org/guidance/ethical_guidance/consent_guidance_common_law.asp#Refusal

GMC Treatment and care towards the end of life: good practice in decision making. 2010.
http://www.gmc-uk.org/guidance/ethical_guidance/end_of_life_care.asp

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. As of April 2015 this work is largely unpublished in peer reviewed journals but is available in final report, recommendations, abstracts and other materials on <http://www.loros.co.uk/education-training-research/research/exploring-mnd-experiences/>

Andersen, PM, Abrahams, S, Borasio, GD, de Carvalho M, Chio A, Van Damme P, Hardiman O, Kollwe K, Morrison KE, Petri S, Pradat PF, Silani V, Tomik B, Wasner M, Weber M. (2012) *The EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis. EFNS guidelines on the Clinical management of amyotrophic lateral Sclerosis (MALS) – a revised report of an EFNS task force.* European Journal of Neurology, 19: 360 - 375.

Ando, H, Chakrabarti, B, Angus, RM, Cousins R, Thornton EW, Young CA. (2014) *Experience of long-term use of non-invasive ventilation in motor neuron disease: an interpretative phenomenological analysis.* BMJ Supportive and Palliative Care, 4: 50 - 6.

Baxter, SK, Baird, WO, Thompson, S, Bianchi, SM, Walters, SJ, Lee, E, Ahmedzai SH, Proctor A, Shaw PJ, McDermott CJ. (2013) *The initiation of non-invasive ventilation for patients with motor neuron disease: Patient and carer perceptions of obstacles and outcomes.* Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 14: 105 – 110.

Baxter, SK, Baird, WO, Thompson, S, Bianchi, SM, Walters, SJ, Lee, E, Ahmedzai, SH, Proctor, A, Shaw, PJ and McDermott, CJ (2013) *The use of non- invasive ventilation at end of life in patients with motor neurone disease: A qualitative exploration of family carer and health professional experiences.* Palliative Medicine, 27: 516 - 523.

Berger, JT (2012) *Pre-emptive use of palliative sedation and amyotrophic lateral sclerosis.* Journal of Pain and Symptom Management, 43: 802 - 805.

Bonner, S, Tremlett, M and Bell, MDD (2009) *Are advance directives legally binding or simply the starting point for discussion of patients' best interests?* British Medical Journal, 339: 1230 - 1234.

Bourke, SC, Tomlinson, M, Williams, TL, Bullock RE, Shaw PJ, Gibson GJ. (2006) *Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial.* Lancet Neurology, 5: 140 - 147.

Borasio, GD and Voltz, R (1998) *Discontinuation of mechanical ventilation in patients with amyotrophic lateral sclerosis.* Journal of Neurology, 245: 717 - 722.

British Broadcasting Corporation (2011) *Inside the ethics committee: Withdrawing Treatment.* Available at: <http://www.bbc.co.uk/programmes/b012r7jn> . Accessed 18th November 2014.

Chan, JD, Treece, PD, Engelberg, RA, Crowley, L, Rubenfeld, GD, Steinberg, KP, Curtis JR. (2004) *Narcotic and benzodiazepine use after withdrawal of life support: Association with time to death?* Chest, 126: 286 – 293.

Christakis, N and Asch, D (1993) *Biases in how physicians choose to withdraw life support*. *Lancet*, 342: 642 – 646.

Clinch A Le B (2011) *Withdrawal of mechanical ventilation in the home: A case report and review of the literature*. *Palliative Medicine*, 25: 378 - 381.

Curtis, JR and Vincent JL (2010) *Ethics and end-of-life care for adults in the intensive care unit*. *Lancet*, 376: 1347 - 1353.

Dreyer, PS, Felding, M, Klitnæs, CS and Lorenzen, CK (2012) *Withdrawal of invasive home mechanical ventilation in patients with advanced amyotrophic lateral sclerosis: ten years of Danish experience*. *Journal of Palliative Medicine*, 15: 205 - 209.

Edwards, M (2005) *Opioids and benzodiazepines appear paradoxically to delay inevitable death after ventilator withdrawal*. *Journal of Palliative Care*, 21: 299 - 302.

Eng, D (2006) *Management Guidelines for motor neurone disease patients on non invasive ventilation at home*. *Palliative Medicine*, 20: 69 - 79.

Faull, C, Rowe-Haynes, C and Oliver, D (2014) *The Issues for Palliative Medicine Doctors Surrounding the Withdrawal of NIV at the Request of a Patient with MND: A Scoping Study*. *BMJ Supportive and Palliative Care*, 4: 43 - 49.

Gannon, C (2005) *A request for hospice admission from hospital to withdraw ventilation*. *Journal of Medical Ethics*, 31: 383 - 384.

Goldblatt, D and Greenlaw, J (1989) *Starting and stopping the ventilator for patients with amyotrophic lateral sclerosis*. *Neurologic Clinics*, 7: 789 - 805.

Heffernan, C, Jenkinson, C, Holmes, T, Macleod, H, Kinnear, W, Oliver D, Leigh N, Among MA. (2006) *Management of respiration in MND/ALS patients: an evidence based review*. *Amyotrophic Lateral Sclerosis*, 7: 5 – 15.

Van der Hoven, B, de Groot, YJ, Thijsse, WJ and Kompanje, EJ (2010) *What to do when a competent ICU patient does not want to live anymore but is dependent on life-sustaining treatment? Experience from The Netherlands*. *Intensive Care Medicine*, 36: 2145 - 2148.

LeBon, B and Fisher, S (2011) *Case report: Maintaining and withdrawing long-term invasive ventilation in a patient with MND/ALS in a home setting*. *Palliative Medicine* 25: 262 - 265.

Lyall, RA, Donaldson, N, Fleming T, Wood C, Newsom-Davis I, Polkey MI, Leigh PN, Moxham J. et al. (2001) *A prospective study of quality of life in ALS patients treated with non-invasive ventilation*. *Neurology*, 57: 153 – 156.

Martin, NH, Landau, S, Janssen, A, Lyall R, Higginson I, Burman R, McCrone P, Sakel M, Ellis CM, Shaw CE, Al-Chalabi A, Leigh PN, Goldstein LH. (2014) *Psychological as well as illness factors influence acceptance of non-invasive ventilation (NIV) and gastrostomy in amyotrophic lateral sclerosis (ALS): a prospective population study*. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 15: 376 - 387.

NICE (July 2010) *NICE Clinical Guideline 105: Motor Neurone Disease; the use of non-invasive ventilation in the management of motor neurone disease.*

Oliver, D (2004) *Ventilation in motor neuron disease: difficult decisions in difficult circumstances.* Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders, 5: 6 - 8.

Oliver, D and Faull C (2013) *Non-invasive ventilation in amyotrophic lateral sclerosis/motor neurone disease.* Minerva Pneumologica, 5: 27 - 38.

O'Neill, CL, Williams, TL, Peel, ET, McDermott, CJ, Shaw, PJ, Gibson, GJ and Bourke SC (2012) *Non-invasive ventilation in motor neuron disease: an update of current UK practice.* Journal of Neurology, Neurosurgery and Psychiatry, 83: 371 - 376.

Oppenheim, S, Bos, C, Heim, P, Menkin E, Porter D. (2010) *Developing Guidelines for Life Support therapy withdrawal in the home.* Journal of Palliative Medicine, 13: 491 - 492.

Phelps, K, Regen, E, Oliver D, McDermott, C, Faull, C. (2015) *Withdrawal of ventilation at the patient's request in MND: a retrospective exploration of the ethical and legal issues that have arisen for doctors in the UK.* (in press)

Piepers, S, van den Berg, JP, Kalmijn, S, van der Pol WL, Wokke JH, Linderman E, van der Berg LH. (2006) *Effect of non-invasive ventilation on survival, quality of life, respiratory function and cognition: a review of the literature.* Amyotrophic Lateral Sclerosis, 7: 195 - 200.

Polkey, MI, Lyall, RA, Davidson, AC, Leigh PN and Moxham J (1999) *Ethical and clinical issues in the use of home non-invasive mechanical ventilation for the palliation of breathlessness in motor neurone disease.* Thorax, 54: 367 - 371.

Rubinfeld, GD (2000) *Withdrawing Life Sustaining Treatment in the Intensive Care Unit.* Respiratory Care, 45: 1399 - 1407.

Szalados, JE (2007) *Discontinuation of mechanical ventilation at end-of-life: the ethical and legal boundaries of physician conduct in termination of life support.* Critical Care Clinics, 23: 317 - 337.

Tweeddale, MG (2002) *Grasping the nettle - what to do when patients withdraw their consent for treatment: (a clinical perspective on the case of Ms B).* Journal of Medical Ethics, 28: 236 -237.

Von Gunten, C and Weissman, DE (2003) *Ventilator withdrawal protocol.* Journal of Palliative Medicine, 6: 773 - 774.

Von Gunten, C and Weissman, DE (2003) *Symptom control for ventilator withdrawal in the dying patient.* Journal of Palliative Medicine, 6:774 - 775.

Welie, JV and Ten Have, HA (2014) *The ethics of forgoing life-sustaining treatment: theoretical considerations and clinical decision making.* Multidisciplinary Respiratory Medicine, 9: 14 -21

The following guidance has informed this document

Guidelines for withdrawing ventilation (NIV) in patients with MND. Leicestershire and Rutland MND Supportive and Palliative Care Group 20th reviewed January 2014.

Guidelines for withdrawing Non-Invasive Ventilation (NIV) at End of Life. University Hospitals of North Staffordshire.

St Wilfrid's Hospice, Chichester. Clinical Guideline No 9b. Withdrawing Non-Invasive Ventilation from MND patients 2009.

Cottrell S, Edwards F, Harrop E, Lapwood S, McNamara-Goodger K, Thompson A. A Care Pathway to Support Extubation with a Children's Palliative Care Framework. Association for Children's Palliative Care 2011.

Consultation

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 information about this list contact:

Association for Palliative Medicine becki@compleat-online.co.uk 01489 565665

Motor Neurone Disease Association

Royal College of Nursing

British Thoracic Society

Association of British Neurologists

[This section to be completed]

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 14b: 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.

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.

e-Training resources:

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.

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.

Resources for professionals and patients relating to advance care planning:

Advance Care Planning: A Guide for Health and Social Care Staff
<http://www.ncpc.org.uk/sites/default/files/AdvanceCarePlanning.pdf>

Advance decisions to refuse treatment: A guide for health and social care professionals 2014.
<http://www.nhs.uk/resources/publications/eolc-adrt.aspx>

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

British Medical Association. Advance decisions and proxy decision-making in medical treatment and research. 2007.

British Medical Association. Assessment of Mental Capacity
<http://bma.org.uk/assessingmentalcapacity>

MND Association: Information sheet 19. Advance Decision to Refuse Treatment
<http://www.mndassociation.org/Resources/MNDA/Life%20with%20MND/information-sheet-19-advance-decision-to-refuse-treatment-adrt.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

Appendix 2.



14B

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:

14A – *Understanding how motor neurone disease (MND) might affect breathing*

14C – *NICE guidelines on non-invasive ventilation (NIV)*

14D – *Troubleshooting when using non-invasive ventilation (NIV)*

14E – *Air Travel and ventilation for motor neurone disease (MND)*

19 – *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 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

Consultation

Appendix 3. 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, and Scotland or Northern Ireland.

1. Legal principles

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 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 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.
5. 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'.
6. 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.
7. 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.
8. 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.
9. Whilst the timing of death may become a medically manipulated variable 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.

10. 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.
11. 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

In 2006 the NHS End of Life Care Strategy published guidance for professionals regarding 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 patients 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 Mental Capacity Act (MCA) for England and Wales (2005) 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:

1. 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.
2. Be able to retain this information and process it.
3. Weigh up the pros and cons and come to a decision.
4. 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 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 decisions are as valid as if the person themselves were 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 to give consent to or refuse treatment him or herself, with appropriate support where		Yes: The person has capacity to make the decision him or herself. The advance decision is not applicable. Ask

	necessary		what s/he wants to do NO: continue with checklist
Is the Advance Decision Valid?			
2	Has the person withdrawn the advance decision? (this can be done verbally or in writing)		YES: This is not a valid advance decision. Make sure that you have identified and recorded the evidence that the person withdrew the advance decision. NO: Continue with check list
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?		YES: 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. NO: Continue with check list
4	Has the person done anything that is clearly inconsistent with the advance decision remaining his/her fixed decision?		YES: 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. NO: The advance decision is valid. Continue with the checklist.
Is the Advance Decision Applicable?			
5	(a) Does the advance decision specify which treatment the person wishes to refuse? (b) Is the treatment in question that specified in the advance decision?		YES: to both (a) and (b): Continue with the checklist NO: This is not an applicable advance decision
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 that the decision whether to refuse treatment needs to be made?		YES: Continue with the checklist NO: This is not an applicable advance decision
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		YES: 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

	anticipated them?		why they would have affected his/her decision had she/he anticipated them, and record your reasoning. NO: Continue with the checklist
Life Sustaining Treatment			
8	Is the decision both valid and applicable according to the criteria set out above?		YES: Continue with the check list NO: This is not a binding advance decision to refuse the specified life sustaining treatment
9	In your opinion is the treatment in question necessary to sustain the person's life?		YES: Continue with the checklist NO: This is a binding advance decision to refuse the specified non-life-sustaining treatment. It must be respected and followed.
10	Does the advance decision contain a statement that it is to apply even if the person's life is at risk?		YES: Continue with the checklist NO: This is not a binding advance decision to refuse the specified life-sustaining treatment.
11	Is the advance decision: <ul style="list-style-type: none"> • In writing AND • Signed by the person making it or by somebody else on his behalf and at his direction AND • Signed by a witness responsible for witnessing the signature, not the decision 		YES TO ALL: This is a binding advance decision to refuse the specified life-sustaining treatment. It must be respected and followed. NO TO ANY: This is not a binding advance decision to refuse the specified life-sustaining treatment

A Best Interest 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. 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 interests decision may need to be made.

A best interests 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 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.

Consultation

Appendix 4. 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 has 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 realize 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 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.

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 in to 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 and legal and 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 lays the foundation for the decision-making and planning of that withdrawal and contributes to good symptom management.

The MNDA have written information available for patients and families (Leaflet 14b - Appendix 2).

Commencing TV is not common in the UK but appears to be an option patients are increasingly aware of and becoming a positive choice made by 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; emphasize 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.
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	Maximize medications used to treat these problems; consider the use of mechanical insufflators/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 familiarize 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 recognize 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 withdrawing 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 LPAHW and 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. In Scotland Electronic Key Information Summaries are enabling this communication and Electronic palliative care coordinating 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 health care plans, statements of advance wishes and ADRT are available and signposted in Appendix 1.

Appendix 5. Specimen Checklist for Assisted Ventilation Withdrawal

Patient details / sticker

One of the following criteria met

Patient with capacity has made a decision to stop assisted ventilation

OR

Patient has a valid and applicable ADRT?

OR

Patient has appointed a LPAHW and a case of best interest is made

OR

Case of best interests made

Affirmed Request

- Senior clinician appointed to coordinate withdrawal
- Discussion with patient and family where, when and how
- Discussion with MDT why, where, when and how
- Contact home ventilation team if not already involved
- Care plan for removal of assisted ventilation agreed

Within 48 hours
of an affirmed
request

At Home

- Contact GP to arrange prescription of symptom control medication
- Equipment: syringes, needles, cannulae

After death

- Collection of equipment and drugs organised
- Audit form completed
- Notify key people about the death

Appendix 6. Audit of Process and Outcomes

It is hoped that this audit will be completed and submitted electronically but the process for this is yet to be finalised.

Background

The purpose of this audit is to provide information that can lead to the improvement of care for patients and their families. We are collecting data about the withdrawal of assisted ventilation at the request of a patient who has any cause for their respiratory failure. This includes patients with:

- Motor Neurone Disease
- COPD
- Duchene Muscular Dystrophy
- Spinal injury
- And other neuro-muscular and lung pathologies

The audit development was funded by the MNDA and led by Professor Christina Faull, a Consultant in Palliative Medicine.

The data will be analysed by the Association for Palliative Medicine and reported as anonymized information that can inform Guidance and practice. Summarized, anonymized benchmarking data will be available to individuals and professional organizations who have endorsed the audit. Non-attributable information may also be submitted for publication in peer reviewed clinical journals.

There is currently very little guidance on withdrawal of non-invasive assisted ventilation (NIV) or invasive assisted ventilation (IV) available for professionals or patients and carers. NICE guidance for NIV in Motor Neurone Disease (2010) states that clarity around the most effective and acceptable method of withdrawal, and how this process should be facilitated and managed, are both needed.

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 utilized 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 should be grateful if you would try and 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 anonymized and all publications non-attributable.

Your name:
Your job/role:
Your email:

Section1: 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)?	Capacity ADRT
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 <16hrs/day 16-22 hrs / day > 22 hrs / 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)	<table border="1"> <tr> <td>Mode of Ventilation</td> <td>Pressure control Pressure support Other</td> </tr> <tr> <td>IPAP</td> <td>cm H2O</td> </tr> <tr> <td>EPAP</td> <td>cm H2O</td> </tr> <tr> <td></td> <td></td> </tr> </table>	Mode of Ventilation	Pressure control Pressure support Other	IPAP	cm H2O	EPAP	cm H2O		
Mode of Ventilation	Pressure control Pressure support Other									
IPAP	cm H2O									
EPAP	cm H2O									
18.	Was the patient on an infusion (syringe driver) before the withdrawal of assisted ventilation was planned?	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?

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 20 & 21 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? Before 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 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 patients 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? After 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:

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

Section 4. After the withdrawal

41.	Was there any immediate feedback from the family about the withdrawal if they were	<p>Yes</p> <p>No</p>
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	present or anything they specifically commented on that may help others to know in the future?	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.

Appendix 7. 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 back up 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 of [Name] still breathing.

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

A qualified professional must attend to verify 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 passed away, 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 quickly.

2) The GP or ambulance crew 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 vent (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.



On/off Switch

Appendix 8. Glossary of Terms and Abbreviations

Advance Care Planning 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 and can relate 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.

BiPAP (bilevel positive airway pressure) provides two levels of airway pressure: inspiratory positive airway pressure (IPAP) and a lower expiratory positive airway pressure (EPAP) for easier exhalation.

IPAP is inspiratory positive airways pressure.

Lasting Power of Attorney (LPA) is a legal tool for a patient to appoint someone to make decisions on their behalf if they lose capacity to do so themselves. 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 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 the upper and lower motor neurones. Degeneration of these neurones leads to weakness and wasting of muscles, causing reduced power in the limbs and difficulties with speech, swallowing and breathing. Eye movements, bladder, bowel and sexual function are generally unaffected. Intellect, memory, sight and hearing are also usually preserved.

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 or nasal mask, or some patients use a combination.

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.

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