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

Guidance for Professionals

Association for Palliative Medicine of
Great Britain and Ireland

November 2024

Endorsements

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

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

Statement of Independence

This guidance does not advocate the use of any particular product.

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Background and Acknowledgments

The Association for Palliative Medicine would like to thank all those who contributed to the updated Guidance. This includes over 63 professionals who have given time and expertise and shared their experiences so openly though submission of a core dataset of almost 100 episodes of withdrawal of ventilation. This pragmatic evidence base has assured that the Guidance enables best care for patients and good outcomes for their families.

The development of the Guidance published in 2015 drew substantially on an interview study with 17 family members and 50 health professionals across the UK and we should also like to acknowledge their enormous contribution to the depth and breadth of this Guidance. Only by exploring what had happened were we been able to understand what was needed and to implement change.

All of this work has funded by LOROS the Leicester, Leicestershire and Rutland Hospice and the Motor Neurone Disease Association. The APM especially acknowledges the leadership of Professor Faull in this 12-year programme of work.

Feedback on this document is welcomed and should be sent to jonathanpalmer@nhs.net

Guidance summary: Principles and Standards for care

The purpose of this Guidance is to support professionals when working with patients with motor neurone disease (MND) who are dependent on their ventilation and ask that this be stopped. The document is structured to provide concise guidance for delivery of safe and effective care, with extensive supporting information included as appendices.

The use of non-invasive assisted ventilation (NIV) improves both quality of life and survival in selected patients with ventilatory failure due to MND. Some people progress to use invasive, tracheal ventilation (TV) which can prolong survival and may maintain quality of life.

For the majority of people NIV use may continue and provide benefit throughout the dying process. Some people using NIV intermittently or just at night may choose not to put it back on and utilise other strategies for symptom management.

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

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

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

- The decision to discontinue assisted ventilation is a unique journey for every patient and their family[§].
- For a patient dying from MND, it is their legal right to decide to refuse assisted ventilation, and the duty of care of professionals to anticipate and manage the physical and emotional impact of this decision on the patient and family members.
- There is a need for discussion and education of the health and care professionals involved in the care of the patient about this ethical and legal context.
- Communication with the patient, the family and between the professionals involved is of fundamental importance in achieving sensitive, safe and effective care.
- Teamwork is key to achieving best outcomes for the patient and requires co-ordination and senior clinical leadership.
- The need for psychological support for the patient, the family and for the professional team should be anticipated and planned for.
- The principles for the management of symptoms are generalisable but the precise methodology requires individual tailoring to the patient.

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.

Timing	Standard	Process
When commencing assisted ventilation and throughout care.	**Standard 1** A patient should be made aware that assisted ventilation is a form of treatment and they can choose to stop it at any time. They should be in no doubt that this is legal and that healthcare teams will support them.	**Page 14** Inform patients that they can choose to stop the treatment at any time, that it is entirely their right and is legal; and that their healthcare team will manage their symptoms in a different way. This conversation should be introduced before assisted ventilation has commenced and the opportunity to discuss further should be actively promoted throughout care thereafter. Offer patients and, with due regard for confidentiality, their families the opportunity to discuss future scenarios when assisted ventilation is being considered. This should include confirmation and reiteration that even if the patient becomes dependent on it that they can still stop it at any time, even if this means they may die soon after. They should know that their symptoms will be managed in a different way and distress minimised. Some may wish for explanation and discussion of dying with ventilation in place. Promote the concept of advance care planning and discussion of wishes and values, especially those for

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[§] In this Guidance the use of the word ‘family’ is inclusive of those close to the patient as well as those actually related.

		whom communication is affected. Include advance decision to refuse treatment (ADRT) and appointment of attorney choices. Assess and discuss capacity for the decision about treatment and its continuation.
Withdrawal of assisted ventilation	**Standard 2** Senior clinicians should validate the patient's decision and lead the withdrawal. **Standard 3** Withdrawal should be undertaken within a reasonable timeframe after a validated request and wherever possible at the patient's preferred location. **Standard 4** Symptoms of breathlessness and distress should be anticipated, pre-empted and effectively managed.	**Page 15** 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 conversation and the patient's final decision. Planning, co-ordination and communication are vital tasks. **Page 16** Communicate and liaise with patient, family and professionals to ensure that the patient's preferred place and time of withdrawal is considered and where possible implemented. Discuss the process of withdrawal, what may happen and how symptoms will be managed. Include in this the potential for living for some hours without the ventilator. Identify key professionals and their roles. This should include a lead for planning care. Ensure members of the team understand the ethical principles and the legal position. **Page 19** Make an individualised plan for symptom management. Good care will require 3 clinicians to undertake the withdrawal. Use opioids (most usually morphine) for breathlessness and benzodiazepines (most usually midazolam) for anxiety. The use and purpose of medication should be explained to family. Those who are **highly ventilator dependent,** and become distressed after very short periods off the ventilator, require: - **anticipatory symptom control** <u>before</u> assisted ventilation withdrawal. - most often **deep sedation** to a level of **unconsciousness** before the ventilation can be withdrawn without causing distressing symptoms.

		- gradual and iterative, step weaning of ventilation support in parallel with testing of level of distress and alertness and titration of medications. Assessing effectiveness of symptom management is an iterative process of reducing ventilation, pausing for a few minutes to observe for symptoms administering further medication to manage symptoms if needed, then further reduction in ventilation in steps to around IPAP 8cm H2O and 8 BPM. Finally removing the mask or tubing only when it is clear that this will not cause immediate symptom distress. Those that are **moderately ventilator dependent** can manage some hours off assisted ventilation are likely to require: - a lower level of anticipatory medication - **milder, conscious sedation** prior to stopping ventilatory support - prompt and ongoing symptom control for emergent signs of distress. Plan for the following: - What drugs, doses, route? - Who will prescribe and administer? - Who will manage the ventilator and how will settings be adjusted? - Who will remove the mask or disconnect tubing? - will the tracheostomy tube be removed?
After death	**Standard 5** After the patient's death, family members should have appropriate support and opportunities to discuss the events with the professionals involved. A care team debrief should be planned.	**Page 32** Consider the needs of family and professionals after death: - Plan who will provide debrief and support to family members. -Plan debrief for professionals/significant event analysis.

The importance of this guidance

A person who is dependent on a ventilation for survival and who decides that they no longer wish to have this has come to a momentous decision. This life-ending decision evolves over time and is incredibly hard to make. Without the ventilator, these people are likely to develop acute and severe breathlessness, so the process of withdrawal needs to be managed in a planned and proactive way. This will ensure that they receive appropriate symptom management and that unnecessary distress is avoided.

Professionals have said that providing the care for a ventilator-dependent patient who has asked for assisted ventilation to be stopped is both practically and emotionally challenging. Lack of guidance on practical aspects of withdrawal, poor advance care planning, lack of experience and the need to support all involved in order to prevent conflict were recurrent themes in research that led to the publication of the 1st edition of the Guidance in 2015. Since its publication audits of 95 withdrawals has shown that the individualised, proportionate and titrated approach to symptom management has led to good outcomes for patients, families and professionals.

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

It seems that around 60% of people with MND use assisted ventilation to manage their symptoms, improve quality of life and, for many, prolong their life. Perhaps 20% of these people who use NIV discontinue it themselves at some point. However, most continue to use it until they are very disabled by the MND and some continue to use it as they die.

A small number of patients who are dependent on assisted ventilation for survival make an elective decision to stop it. 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. Our dataset indicates that a minority of patients make a written statement or directive with respect to withdrawal in advance of their losing the ability to communicate or losing capacity for another reason and that even fewer appoint an attorney for decisions about life-sustaining treatments. The importance of clinicians documenting discussions about wishes and values is therefore pivotal.

Evidence suggests that not all patients know about their potential choices, what dying is like when using assisted ventilation, or are asked about their views of continuing assisted ventilation. There is a clear need for more information sharing and improvement in facilitated decision making and there is now evidence that making proactive decisions and well planned ventilation withdrawal can be empowering for patients and cherished by families.

The aim of the Guidance is first and foremost to improve the care of patients and families. The application of the Guidance should also lessen the emotional impact on professionals.

We recognise that whilst primarily aimed at professionals some of the content may also be useful to patients, families and others in supporting people with MND including those. The Guidance will also be useful in supporting the care of people with MND where capacity has been lost and where *best interests* decision-making is required and for people with other neuro-muscular conditions.

Part 1: The context

An overview of assisted ventilation in MND

MND (also known as amyotrophic lateral sclerosis, ALS) is a progressive, invariably adult onset, neurodegenerative disease for which there is no highly effective disease-slowing therapy. People vary in the way MND symptoms first emerge, the pattern of progressive weakness of muscles arising from the loss of motor neurons of the brain and spinal cord, and the speed of disability progression. However, at some point, almost all those with MND develop weakness of respiratory muscles. The most frequent cause of death in MND is respiratory failure secondary to impairment of the respiratory musculature, and occurs within 2-3 years of the onset of first symptoms in a majority of cases.

Non-invasive ventilation (NIV) is a medical treatment that can improve quality of life, symptoms and survival in selected patients. Guidance from the National Institute for Health and Care Excellence (NICE) supports its use. Undoubtedly, NIV has many benefits for patients and the majority of patients choose to at least try NIV. In the UK the use of NIV has increased markedly over the past 10 years. It is thought that around 60% of patients with MND opt for a trial on NIV with maybe 80% of people successfully adapting to ongoing use of NIV.

A very small number of patients may choose assisted ventilation via tracheostomy (TV) if NIV is not tolerated or does not support ventilation effectively. Some have TV initiated in an unplanned way when presenting in crisis. It is estimated that no more than 1% of patients with MND in the UK have assisted ventilation via tracheostomy.

Decision-making around starting NIV is complex. People 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 making the decision about starting the treatment. It is of great importance that the patient considering NIV is aware that they can discontinue NIV at any stage in the future if this is their wish.

To start with most people use NIV for discrete periods of time, most often at night only. Many people progress to using NIV for much longer periods of the day and eventually become very dependent, unable to tolerate even a few minutes without it (e.g. for cleaning teeth or drinking). A small number who are dependent on NIV for survival use NIV mainly at night, with only short periods of use in the daytime, additional management options to palliative breathlessness are used by these patients to manage symptoms of severe breathlessness by day. Most patients on TV will progress to using this 24hours a day and some, but not all, will be unable to make any respiratory effort themselves.

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

It is not possible to predict which people might follow this route when treatment is commenced.

Experiences of decision-making and withdrawal of assisted ventilation

Research has explored the experiences of people thinking about and making the decision to stop their assisted ventilation. In summary this has found that:

- Many reported having limited understanding of the withdrawal process (and of dying with the ventilator in place).
- People who made proactive decision about withdrawal felt this allowed them some control over the time and place of their death.
- A significant proportion do not wish to actively make advance plans. Sometimes this was due to a lack of understanding. Sometimes it was linked to an acknowledgement of their future change in abilities or health events, and how this might affect their quality of life – so they wanted to make decisions in timely congruence with this.
- Those who do not wish to talk about their own end life, can be encouraged to discuss the concepts to allow families and HCPs to make well informed 'best interests' decisions if needed.
- Decisions to withdraw ventilation are often relational and evolving.
- People may want to consider achieving or missing specific dates, seasons or other factors that impact those left behind.

While many family members reflect positively on the withdrawal of ventilation as achieving what their loved-one wanted many indicate experiences that could be improved. Evidence indicates that:

- The role of the family in advocating for the patient and achieving their wishes can be a very heavy burden.

- There may be conflict within the family, which needs to be managed.

- There is insufficient information given to patients and families about choices and reassurances about the legal right to stop assisted ventilation.

- More opportunities to discuss concerns and options should be offered.

- Once they have expressed their choice to withdraw treatment, delays in carrying out the patient's wishes are distressing for patients and their families.

- Families need information about what will be done in the withdrawal process, how long it might take for the patient to die and any symptoms or changes they may expect to see (e.g. gasping, colour change).

- Families are frequently asked or even expected to take a role in the actual withdrawal of the ventilator and are sometimes left alone during the process.

- Families are supported by teams who are inexperienced and sometimes unconfident. This can lead to poor outcomes which families felt could have been prevented by experienced specialist input.

- If a patient experiences distressing symptoms during withdrawal, this has a significant effect on the relatives present. Relatives' perceptions of distressing symptoms may be different to those of the healthcare professionals involved.

- Family members can feel very isolated and unable to discuss the situation before or after with their usual social support network.

- Relatives may need additional support after the withdrawal and still have questions and issues they need to discuss with the healthcare professionals involved.
- Some family members were left with feelings of guilt or shame and did not discuss the patient's death with friends for fear of being mis-judged as assisting suicide.
- Cases where the withdrawal does not proceed as expected (e.g. delays in enacting the decision, symptoms not controlled, withdrawal requiring several attempts, relative being required to play a more active role than anticipated) are more likely to leave relatives with negative feelings about the withdrawal.

Challenges in the withdrawal of assisted ventilation

Interviews with health professionals and family members about their experiences in withdrawing assisted ventilation at the request of a ventilator-dependent patient with MND identified key challenges. The collection of data since the Guidance was published in 2015 affirms these as:

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

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

The evidence base for symptom management during withdrawal of assisted ventilation

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 a small volume of literature discussing the clinical and practical aspects of withdrawal of assisted ventilation in MND and a little on the withdrawal of assisted ventilation for other conscious patients. We have now collated the information from 68 professionals from across the UK who have undertaken the care of 95 people with MND from across England who had their ventilation withdrawn. The majority were doctors working in palliative medicine (n=56). Other healthcare professionals who submitted proformas were physiotherapists, speech and language therapists, staff nurses, and clinical nurse specialists in home ventilation, palliative medicine, respiratory medicine, and neurology. This data set has been published in part (Faull and Wenzel 2022) and we hope for further publication in 2024-25.

In the data set we found that:

- most people used the SC route but the IV was also frequently used.
- 94% of patients received at least one dose of medication immediately prior to stopping the ventilation (mode=1 dose; range=1-8 doses).
- 44% of patients required no medication after the ventilation was stopped,
- Those requiring further medication mostly required 1 further dose of midazolam and/or opioid but a few required more (range =1-5 doses).
- 10% of patients needed the addition of levomepromazine as a sedative.

The mode and range of doses of midazolam and morphine equivalent pre- and post-withdrawal are shown below (Table 1).

Table1: Medication use in the withdrawal of ventilation

	Pre-withdrawal (n=89)	**Post-withdrawal (n=52)**	**Total (n=94)****
Midazolam			
Median (mg)	15	10	20
Mode (mg)	10	10	20
Range (mg)	2-120	0.5-30	0.5-120
Opioid*			
Median (mg)	15	10	20
Mode (mg)	5	5	10
Range (mg)	2-55	5-40	4-60

**opioid doses given as the subcutaneous morphine equivalent.*

***n=94 as one patient did not receive any medication due to withdrawal being patient initiated and unplanned.*

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, not always predictable and often unexpected by patients, families and the professional team.

In the data set of 95 patients, we found that:

- the most common time to death after mask/tubing removal was 15 minutes.
- 69% of patients had died within 1 hour.
- the time to death ranged between 1 minute and 54 hours.

The impact of withdrawal of assisted ventilation on health professionals

Stopping something that has been keeping a person alive is a difficult situation for all concerned. The withdrawal of life-sustaining treatment from insentient patients who are dying from organ failure or brain injury (not MND) in intensive care units has an emotional impact on professional team members, and physicians consider mechanical ventilation the most difficult treatment to withdraw. In the context of MND where patients may remain able to hear, see, think and feel normally, but may not be able to communicate, the impact on their professionals could be expected to be even greater.

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

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

> *79. You can withdraw from providing care if your religious, moral or other personal beliefs about providing life-prolonging treatment lead you to object to complying with: (a) a patient's decision to refuse such treatment, or (b) a decision that providing such treatment is not of overall benefit to a patient who lacks capacity to decide.*
>
> *However, you must not do so without first ensuring that arrangements have been made for another doctor to take over your role. It is not acceptable to withdraw from a patient's care if this would leave the patient or colleagues with nowhere to turn.*
>
> *(GMC 2022)*

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

There is indication then, that not only does the consideration of, or actual withdrawal of, assisted ventilation have potential for significant and extraordinary impact on healthcare professionals, it may also have a direct effect on their practice. To achieve best outcomes for patients and their families, the impact on professionals and the support they require needs to be anticipated and planned for.

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

1. Sharing information and discussing choices

Standard 1

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

When breathing becomes impaired and assisted ventilation is being considered, patients and, with due regard for confidentiality, families should be offered information (verbal and written) and the opportunity to discuss and ask questions about the benefits and burdens of ventilator support. Some discussion about potential future choices and scenarios, including ventilator dependence may also be important for patients (see Appendix 2)**.** Some may want this, others may not. This sort of discussion might include:

- Explanation of what happens as breathing becomes worse.
- Discussion of dying with ventilation in place.
- Confirmation and reiteration that even if the patient becomes dependent on ventilation, it that they can still stop it at any time. It's legal and their right to stop it.
- That stopping ventilation when they have become dependent means they may die soon after.
- Management of symptoms in a different way and minimising distress.

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

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

> '*You can stop using this treatment at any time you want to. If you are using it a lot you may want some help to manage any problems such as increased breathlessness that may occur when you stop it. You can ask to stop this even if you are completely dependent on a ventilator to help you breath*. We would then find another way, usually with medications, to manage the symptoms you have.'

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

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

The ethical and legal principles and their application to decision making are discussed in Appendix 1.

2. Deciding and planning the withdrawal

Standard 2
Senior clinicians should validate the patient's decision and lead the withdrawal.

A senior doctor should take responsibility for validating the decision to withdraw assisted ventilation. A senior health care professional (not necessarily a doctor) should take responsibility to co-ordinate the process and the team. Communication with the patient, family members and the broad professional team is a key component of high quality care. There may be key things that the patient wants to put in place before they proceed.

Patients make settled decisions about the withdrawal of assisted ventilation over time and many factors support and influence this. The journey towards the decision to withdraw assisted ventilation and the role of professionals in this journey is discussed in detail in Appendix 2.

Although some patients will have been considering withdrawal for some time, the point at which they will make a final decision that the burdens of assisted ventilation outweigh its benefits, is unpredictable and often one of immediacy ['I want this to stop now']. In practice, however some patients will identify a future time ['I want this to stop after Christmas'].

People with MND can donate their organs (but not eye tissue). This will require a specific type of planning and withdrawal of ventilation will take place in the operating theatre environment with organ retrieval soon after.

Validating the decision

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

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

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

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

See Appendices 1 and 2 for additional guidance.

Standard 3
Withdrawal should be undertaken within a reasonable timeframe after a validated request and wherever possible at the patient's preferred location.

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

Planning and co-ordination

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

The co-ordinator may be the:

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

What to plan for

Who:

- needs to be informed about the planned withdrawal (MDT, family, paid carers, other community care staff)?
- will manage the ventilator?
- will remove the mask/tubing?
- will ensure adequate supplies of medication, syringes and other supplies are available? Particularly important If the withdrawal is happening at home.
- will prescribe and or administer the medication? A GP would prescribe the medications for a patient at home.
- will have the key role of supporting the family?
- will confirm that the patient has died and inform professional team members?
- will complete the medial certificate of cause of death?

A minimum of three people are required to ensure a withdrawal is managed effectively, 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. An MDT approach is advocated. Symptom management is likely to be led by a doctor but another advanced practitioner with appropriate prescribing rights could lead the withdrawal. One of the team needs competence in managing the ventilator settings.

Where will it take place?

Wherever possible the co-ordinator should plan to deliver the withdrawal at the patient's preferred place. However, the patient's preference may not be feasible particularly if withdrawal is required in a timely fashion or family views are not congruent with this. Consider:

- What are the anticipated challenges for the preferred place and how can they be overcome?
- Is there a difference in preference for the patient and family, and what problems does this cause for the family?
- Has the patient expressed a desire to donate organs?

When will it take place?

- Provision for professional continuity of support for care and symptom management will need to be planned for several hours and sometimes needed for up to 2 to 3 days.
- Co-ordination of professionals' availability and family support (especially if family travelling to be there).

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

- What drugs will be used?
- How will drugs be delivered?
- Is there a separate room to use for drawing up medication and for staff to be in to give the patient and family privacy?
- What other equipment will be required and how will it be organised?
- How will mask/tube and ventilator be managed?
- Are there clear instructions as to how to turn the ventilator off/ change settings/stop alarms?
- For a patient using TV, preferences for what to do with the tracheal tube (remove or leave in situ) should be explored.
- Has a DNACPR/allow natural death form been completed?

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 the use of this time to say goodbye. Where possible withdrawal should start early in the working day.
- Symptom control – including likely symptoms, what medications will be given, how they will be given, what the objectives are including level of sedation and patients' preferences.

- Mechanics of withdrawal including who will do what and that the level of symptom control will be tested by reducing ventilation, assessing response and considering options to control any symptoms usually with medication, to ensure all is well before finally stopping.
- What will happen once the mask/ventilation has been removed? Acknowledge the uncertainty about rate of deterioration to death on stopping assisted ventilation. Allay expectations that this will be immediate and explain it could be hours (and longer in exceptional circumstances). Address fears about distress and what would happen should distress be evident.
- Discuss what additional professional support may be required until death occurs.
- What will happen after they die?

Who would the patient ideally like to be present?

Advise about the healthcare professionals who will need to be present and discuss their wishes about the presence of friends, family and the paid carers and health professionals that have supported them in the community. Spiritual support and religious rituals may be of significance for the patient. The availability of a nearby room for family is important as often they need some space for ‘time out’.

What to discuss with patient and family

The elements of the discussion with the patient should, with due respect for confidentiality, also be had with the family. This should ideally take place at the same time unless this is not desired by the parties concerned. The difficulty and impact of the decision should be acknowledged, and concerns and expectations explored. Several members of the MDT, including chaplaincy, can support these discussions.

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

Additional points for discussion with the family

- What, if any, role might they wish to have? Some family members do want to remove the mask and turn off the ventilator, many will not.
- Ensure that family members and more junior team members are aware of the changes that might occur close to death and that these do not signify distress. For example some can become very dark blue, there can be eye opening despite deep sedation and/or agonal breathing.
- Discuss what family may want from professional presence (maybe in separate room?) if there is a long time after withdrawal.

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

<u>What to discuss with other professionals</u>

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. There can be a very extensive group of people particularly if the withdrawal takes place in the patient’s home and if have had an extensive team of paid carers and primary care professionals.

The lead doctor and/or the named co-ordinator should discuss with all those who have been/will be caring for the patient some of the following aspects:

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

3. Undertaking the withdrawal and symptom management

Standard 4

Symptoms of breathlessness and distress should be anticipated, pre-empted and effectively managed.

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

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

Principles for symptom management

A minimum of three people are required, at least at the start of the withdrawal process:

- one to manage the ventilator.
- one to manage the symptom management with medication adjustment.
- one to focus on supporting the family.

An MDT approached is advocated. Symptom management is likely to be led by a doctor but another advanced practitioner with appropriate prescribing rights could lead the withdrawal. One of the team needs competence in managing the ventilator settings.

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

- How quickly the patient becomes distressed without assisted ventilation.
- Choice of drugs.
- What drugs the patient is already on.
- Which route of administration the clinical team feel confident with.
- Who is administering medication?
- The preferences of the patient and family.
- The ease of venous cannulation.

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

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

High ventilator dependence

> **This group comprises those patients who become very breathless or distressed within minutes of not having this in place. These patients will usually require deep 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 iteratively reducing ventilation settings in small steps and adjustment of medication as necessary as described in **Managing the ventilator** and **Medications** section below.

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 stimuli and on the 'test' reductions of the assisted ventilation, no symptoms are precipitated.

Moderate ventilator dependence

> **This group refers to those patients who can tolerate longer periods of time without assisted ventilation and who will not immediately develop symptoms without ventilation.**

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.

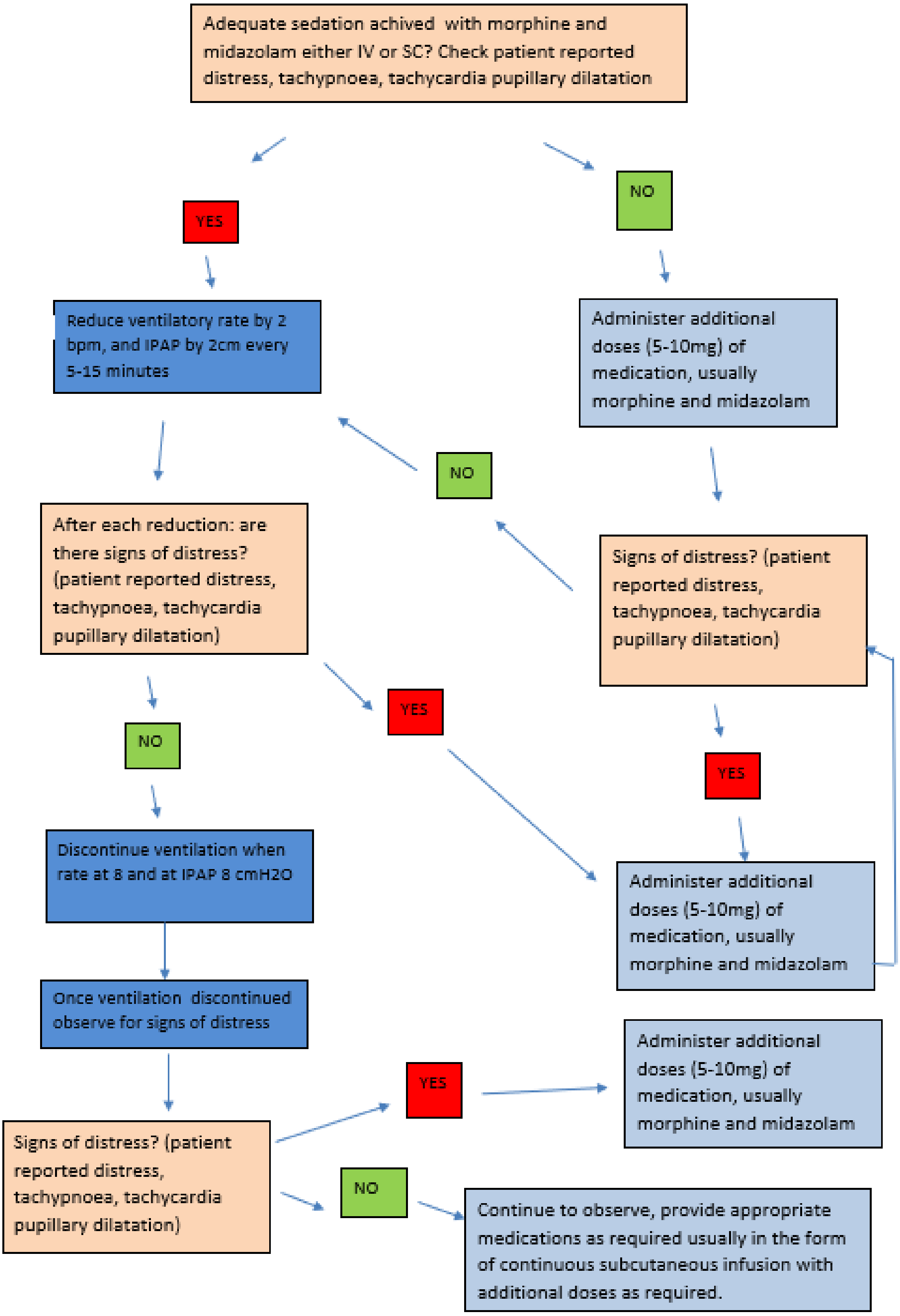
Managing the ventilator and symptoms

Withdrawal of ventilation in patients who are dependent on ventilation can present a significant challenge in terms of symptomatic and ventilator management. It is therefore recommended that the specialist home mechanical ventilation team be present during the withdrawal.

If the patient's specialist home mechanical ventilation team cannot be present at the withdrawal, they must be contacted as part of the coordination and planning of the withdrawal. The mode of ventilation along with details on how to unlock the device, manage alarms, alter parameters and turn the device on and off must be known before the withdrawal commences.

Ventilated patients with neuromuscular disease such as MND are dependent symptomatically on adequate tidal volumes. To avoid any distress it is essential therefore, that the ventilator settings are reduced gradually (weaning) with adjustment of medications to manage symptoms as needed in an iterative process (see figure 1).

Figure 1. Managing the ventilator and symptoms.



Before reducing settings, all alarms that can be switched off must be disabled. On some devices, not all alarms can be switched off and therefore alarm volume levels must be reduced to the lowest level. External heated humidification devices can also alarm, these devices should be switched off before ventilator settings are reduced.

During this stepped weaning process, if the patient demonstrates any signs of distress, additional doses of medication, usually (5-10mg SC) morphine and midazolam, should be administered (see medication section below) and sufficient time given for this to work before any further reduction of ventilator settings.

Provided that sedation is adequate and the ventilation is reduced slowly, it is unlikely that the ventilator settings will need to be increased or ventilation recommenced.

Steps in reducing ventilation.

To avoid the likelihood of distress as the ventilation is reduced, it is recommended that both the ventilator rate and pressures are reduced together in steps. As a guide:

- Reduce back-up respiratory rate in 2bpm (breaths per minute) steps, every 5-15 minute to 8bpm.
- Reduce inspiratory pressure (IPAP) in 2cm H2O steps, every 5-15 minutes to 8cm H2O.

Once the rate has been reduced to 8bpm and the IPAP to 8cm H2O, and the adequacy of sedation is appropriate the ventilator can be disconnected.

If the patient shows signs of distress after removal of ventilation additional doses (5-10mg SC) of medication, usually morphine and midazolam, should be administered as required.

A small number of patients will have a less common mode of ventilation and as well as needing reductions in respiratory rate pressure, volumes will also need to be reduced. In patients using these ventilators it is recommended that in parallel to reducing the rate and inspiratory pressures (IPAP MAX and IPAP MIN Pressure), that the tidal volume is reduced by 50-100mls every 10-15 minutes to 300mls.

Medication

Route of administration

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

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

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

No other routes of drug administration are recommended.

Agents

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

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

Benzodiazepine: midazolam is the drug of choice

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

In rare instances phenobarbitone and propofol have been needed for sedation

Doses

The dose of medication that patients require to manage symptoms is quite variable and is not greatly predictable on the level of ventilator dependence or prior use of opioids or benzodiazepines.

However, the total doses of medication in our dataset grouped by age suggest that younger patients were more likely to require higher doses of medication, particularly before withdrawal.

BEFORE WITHDRAWAL	**Age 30-50** (n=12)	**Age 51-70** (n=53)	**Age >70** (n=29)
Midazolam			
Median (mg)	30	15	15
Mode (mg)	30	10	10
Range (mg)	10-120	5-55	2-45
Frequency (n)	11	47	18
Opioid*			
Median (mg)	25	15	15
Mode (mg)	40	5	5
Range (mg)	5-45	5-55	2-47.5
Frequency (n)	11	45	25

**opioid doses given as the subcutaneous morphine equivalent.*

AFTER WITHDRAWAL	**Age 30-50** (n=12)	**Age 51-70** (n=53)	**Age >70** (n=29)
Midazolam			
Median (mg)	10	10	6.5
Mode (mg)	10	10	10
Range (mg)	10-20	5-30	0.5-30
Frequency (n)	6	26	17
Opioid*			
Median (mg)	10	10	7
Mode (mg)	10	5	5
Range (mg)	5-30	5-40	2-20
Frequency (n)	6	21	12

**opioid doses given as the subcutaneous morphine equivalent.*

TOTAL	**Age 30-50** (n=12)	**Age 51-70** (n=53)	**Age >70** (n=29)
Midazolam			
Median (mg)	40	20	20
Mode (mg)	40	10	20
Range (mg)	10-120	10-60	0.5-55

Frequency (n)	11	49	28
Opioid* Median (mg) Mode (mg) Range (mg) Frequency (n)	30 30 7.5-60 11	20 10 7.5-60 47	20 5 4-47.5 25

**opioid doses given as the subcutaneous morphine equivalent.*

The doses below are appropriate starting points. It is vital to titrate medication to effective symptom management. The appropriate dose will vary with age and physiological resilience, and other reasons including the medications already in use for management of the patient's symptoms.

Many patients are already on continuous SC infusion of medications to help manage symptoms. For some patients this is increased, or an infusion commenced, to manage anxiety or breathlessness a few hours before the withdrawal of ventilation is commenced (see case examples in section 4).

To achieve deep, unconscious sedation for highly ventilator dependent opioid naïve[**] patients:

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

The period of time between the anticipatory administration of medication and the final removal of assisted ventilation is influenced by the route of drug administration and by independent patient variables. Sufficient time must be allowed for stepped ventilator weaning to ensure that the patient has an adequate level of sedation before assisted ventilation is withdrawn. For IV administration, this may require only minutes of assessment and titration of medication before ventilation withdrawal (mode =15 minutes, range 8 minutes to 2.5hrs in the data set). For SC administration, this may be considerably longer (mode =30 minutes, range is 5minutes to 18 hours) and this should be factored into the care plan.

To achieve mild, conscious sedation for opioid naïve patients[††] who are moderately ventilator dependent.

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

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

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

The timing of giving the initial medication is related to the patient's degree of ventilation impairment, the effect of any SC infusion recently commenced, and the route of administration of the drug. Most but not all patients will require a stat dose of SC medication 20-30 minutes before mask removal.

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

Other aspects of symptom management

Oxygen

This does not appear to be required for symptom management.

Position

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

4. Case examples

These are anonymized, real-world examples from our dataset of 95 patients to give insight into what care may look like. They are not all 'perfect', but intentionally illustrate variability in patient need, the challenges that arise and how the clinical approach needs to be responsive to this. In reality, each of the clinicians reflected on areas they would probably do differently 'next time' in the technical aspects of withdrawal. The ventilator in some cases may have be managed differently to the guidance detailed in the *Managing the ventilator and symptoms* section above. This is because the updated guidance is based upon the audit data analysis and subsequent expert consensus.

Examples of the decision-making process

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

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

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

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

PQ had lost all communication. His ADRT developed with his GP stated that he would want to stop his TV when he lost communication. The GP assessed his ADRT as valid and applicable. The palliative care consultant who had not met PQ before visited and assessed PQ and the ADRT. His wife and family raised no concerns that PQ had changed his mind at any point since making his ADRT. Withdrawal of ventilation was planned with the family for a few days later at home.

DLG, a man in his early 50s was diagnosed with MND 15 years earlier. He started on NIV almost 4 years after diagnosis. He tolerated this for a year or so before being transferred to TV following an acute respiratory problem. DLG had a young family and would always say he wanted to live for as long as possible irrespective of disability. He would not engage in end-of-life discussions preferring to concentrate on "taking each day as it comes". He lived with TV for 8 years and in discussions with him and his wife he continued to express a view that he wanted to live as long as possible and that he would never want to withdraw. He eventually agreed to have a 'DNAR' order completed and agreed he would not want hospitalisation. After 6-7 years on ventilation, it was noted that DLG's communication was worsening, he progressed from eye gaze to e-tran frame, eventually being unable to answer yes and no. During periods of infection, he would often lose the ability to communicate at all. When he recovered from these and was asked about the future, he would still state he wanted to be alive and did not want to withdraw. He refused to draw up an ADRT. His wife had Lasting Power of Attorney for property and financial affairs but not for health and well-being. As time progressed his ability to answer yes and no consistently worsened and the MDT could not accurately determine whether he wanted to continue ventilation. Discussions with his wife happened on several occasions and by a number of professionals. His wife would always refer to DLG's previously expressed wishes. She felt he was not distressed and continued to enjoy watching sport on the television and being around his family. A few months later DLG developed a chest infection and he was not able to communicate at all, this did not change. A respiratory physician attended and discussed the situation with the wife and eldest daughter. His wife and daughter felt that his quality of life was now very poor and it was agreed by the team; respiratory physician, home ventilation team, GP and palliative care that it was no longer appropriate to continue ventilation and it was therefore in his best interests to withdraw. A withdrawal at home was arranged at home a few weeks later.

An example of a patient who chose admission to the hospice to withdraw from mask ventilation where the intravenous route was utilised.

SS was a man who started non-invasive ventilation to manage respiratory muscle weakness 9 months ago. He was now using mask ventilation 24 hour per day and could manage just a few minutes off the ventilator before becoming distressed. He was able to communicate verbally and was fully dependent on others for all aspects of his care. He had capacity and was able to express his desire to withdraw ventilation to a number of health care professionals. His view was settled and discussed with his GP, his neurologist and a palliative care physician.

He asked to be admitted to the hospice to be withdrawn. Despite NIV at high pressure (26cm H2O IPAP, 4cm H2O EPAP) and morphine sulphate slow release 10mg BD, he was still experiencing breathlessness in the days before admission. Upon admission he continued to express his desire to stop treatment and this was planned for the next morning. He was not given additional medication the night before the procedure and the ventilator settings were not altered.
The next morning, he confirmed that he still wanted to withdraw. An intravenous cannula was inserted and the withdrawal commenced.

All times in minutes.

Time zero: 10mg each morphine and midazolam, intravenously.

Time zero to T15: ventilator settings slowly reduced to 50% of patient's norm.

T15: SS looked settled, he did not respond to voice or touch. To ensure his continued comfort a further 5mg of morphine sulphate was given intravenously prior to removing the mask and stopping ventilation.

T20: ventilation stopped and mask removed.

T25: light leg movements and increased work of breathing noted, which could have indicated some distress. Further 10 mg intravenous midazolam given.

T35: SS died peacefully.

An example of withdrawal of TV using the SC route for anticipatory symptom management

JC was a 65-year-old lady who had been dependent on ventilation via tracheostomy for a year or so. She had an ADRT that stipulated that she would want ventilation withdrawn once she was unable to communicate. Withdrawal in her best interests based on her ADRT was agreed by the MDT, (palliative care physician, GP, home ventilation team) and the patient's family. She was very disabled by MND. In the weeks leading up to the withdrawal she was bed bound and appeared to become more distressed. A syringe driver of morphine and was therefore commenced. This required titration to morphine 15mg/24hr and midazolam 15mg/ 24 hours after 24 hours. The withdrawal was commenced 48 hours later and was led by a consultant palliative care physician with the home ventilation team and district nursing team present.

All times in minutes.

Time zero: 5mg each morphine and midazolam, sub-cutaneously.

T0 – T30ventilator BUR reduced to 8, ventilator pressures left the same.

T30: increased work of breathing and in particular increased respiratory rate indicated there may be some distress therefore 10mg each morphine and midazolam, sub-cutaneously.

T60: pressures reduced to minimum and BUR down to 5, JS was comfortable and ventilation was stopped.

T65: JC started frowning therefore further 5mg morphine and midazolam sub-cutaneously.

T85: JC died peacefully.

An example of withdrawal of mask ventilation using the SC route at home

SG was a 75 year old woman who had been dependent on ventilation via mask for a year or so. She was using the NIV for almost 24 hours per day and could manage just a few minutes off. She was highly disabled and relied on others for all of her care. The ventilator settings were IPAP 14, EPAP 5, respiratory back up rate (BUR) 15bpm. She could communicate verbally and had capacity. She expressed that she wanted to withdraw from ventilation at home. The decision was confirmed by a number of professionals including the palliative care physician, neurologist and the home vent team. She felt breathless and was anxious despite the NIV.

On the morning of the planned withdrawal, she confirmed her wish to withdraw to the palliative care consultant; a palliative care physio and home ventilation team specialist nurse were also present.

All times in minutes.

Time Zero: subcutaneous administration of Midazolam 15mg and Morphine 10mg.

T30: further subcutaneous administration of Midazolam 10mg and Morphine 10mg as SG was still too aware to withdraw ventilation.

T55: following titration of ventilator settings down to IPAP 10, EPAP 5 and BUR 8bpm, ventilator was removed. No further medications were required.

T80: SG died peacefully.

An example of withdrawal of TV using the IV route for anticipatory symptom management

JS was 60 when she was diagnosed with MND, she started NIV 12 months later and quickly progressed to tracheostomy ventilation. She was entirely dependent on tracheostomy ventilation for 2 years before asking for the ventilator to be withdrawn.

She was admitted to the hospice 5 days later. Upon admission she was completely dependent on others for all aspects of her care and was able to communicate slowly using an eye gaze system.

She was clear upon arrival that she still wanted to withdraw, her family were aware and although they agreed with her decision, they were finding it difficult to understand.

The morning after admission her view was settled and the withdrawal commenced, it was led by the consultant palliative care physician, the home vent team and hospice nurses were also present. A cannula was inserted.

All times in minutes.

Time Zero 10mg each morphine and midazolam, intravenously.

T10: not adequately sedated, still aware so further 10mg midazolam given, first decrease in ventilator settings by 2cm IPAP and 2 bpm. Alarms switched off.

T15-T25: Very settled, IPAP and BUR reduced to 8cm H_2O and 8bpm respectively.

T25: ventilation stopped and removed. Tracheostomy tube left in place.

T51: JS died peacefully.

Tracheostomy tube removed as part of ‘last offices’.

An example of withdrawal of mask ventilation where high doses of opioids and benzodiazepines were required.

TWP was a 31-year-old man with MND diagnosed in 2014. He commenced Non-Invasive Ventilation in 2016 to manage symptoms of nocturnal hypoventilation. He became dependent on the ventilator following a hospital admission with pneumonia a few months later. Following discharge from hospital, symptoms of breathlessness and distress were managed with lorazepam 0.5 mg BD and 1 mg ON. He was also taking Zomorph 20 mg ON and Oramorph 5 mg QDS PRN.

He requested withdrawal of non-invasive ventilation. This was arranged 11 days after the request but at a time convenient to him and his family. 24 hours prior to the planned withdrawal, his enteral benzodiazepine and opiate was changed to a subcutaneous infusion, morphine 30 mg and midazolam 40 mg over 24 hours. At this point he was using NIV with IPAP of 26cmH_2O and an EPAP of 3cm H_2O, backup rate 15 breaths per minute.

On the day of withdrawal, he remained fully conscious with capacity and the clinical teams including the home vent team and palliative care team were able to confirm his previously expressed wishes. He was given a sub-cutaneous bolus of 10mg each morphine and midazolam. After this ventilation was gradually reduced. Initially, the breaths per minute were reduced by 1-2 at a time down to 8 breaths per minute. Following this, the ventilatory pressures were reduced by 2 cm H_2O at a time down to 12/3. Between changes in ventilation, at least 20 minutes were allowed to assess any signs of distress. TWP required a significant number of boluses to manage symptoms. The overall period from commencing the withdrawal process to withdrawal of mechanical ventilation was 7 hours. During this time, he received a further 2 X 10mg boluses of morphine subcutaneously and 9 X 10 mg boluses of midazolam subcutaneously.

Following withdrawal of mechanical ventilation, he continued to breathe for a further 2 hours before dying without any sign of distress.

An example of withdrawal from a patient who was less dependent on NIV.

SPM was a 60 year old man with MND who was using NIV all night and for periods during the daytime. He had used NIV for 9 months and could manage a few hours without the device. He could communicate verbally, was still able to walk and use his hands to undertake some tasks. He was admitted to the hospice as he wanted to stop using ventilation. He was fully alert and was not particularly anxious or breathlessness upon admission. He confirmed he wanted to withdraw and it was agreed to manage symptoms preemptively and to not start NIV that evening.

All times in hours

T zero: stat dose of 5mg midazolam and 5mg Morphine directly prior to being started on a sub-cutaneous syringe driver containing 10mg midazolam and 10mg Morphine (over 24 hours) 3 hours before the NIV was normally due to be started that day.

T1.5: developed a headache and dyspnoea, therefore stat dose of 5mg midazolam and 5mg Morphine given.

T6: complaining of headache and dyspnoea, looked anxious therefore stat dose of 5mg midazolam and 5mg Morphine given and syringe driver increased to 30mg midazolam, 30mg Morphine and 25mg levomepromazine.

T8: complaining of headache and dyspnoea, looked anxious, therefore stat dose of 5mg midazolam and 5mg Morphine given.

T9: very breathless and restless, but quite sleepy, therefore stat dose of 5mg midazolam and 5mg Morphine given.

T9.5: died peacefully.

An example of a patient who used ventilation overnight who was exceptionally breathless by day but who rarely chose to use NIV by day.

FD was a man who started NIV to manage symptoms of sleep disordered breathing and orthopnoea. He was also in mild daytime ventilatory failure, (VC 50%, ABG: late pm 6.5kpa, HCO3 33.0mmols). He was clear he was not using it for the life prolonging effects.

Three months after starting NIV he asked for it to be stopped. He was using NIV for 9 hours each night, he rarely used it by day despite being exceptionally tachypnoeic with respiratory rate of 35-40 per minute. Although, he was not dependent on NIV, it was clear to his health care professionals that without NIV he would have died weeks or months ago.

Upon admission to the hospice FD was able to transfer from bed to chair with minimal assistance and could use a hand-held call bell to summon help. He had minimal changes to his speech and had capacity. He was fully aware of the implications that withdrawing NIV would bring about his end of life. Because he was so short of breath whilst lying down NIV had to be used when he was in bed. It could not be stopped without causing considerable distress overnight, when out of hours cover would be minimal and symptoms hard to manage. He was very anxious and chose to be admitted to the hospice.

The night before the 'withdrawal' his NIV was commenced at bedtime, a 24hr SC infusion was set up containing morphine 10mg, midazolam 10mg and levomepromazine 10mg. Mask ventilation was started in the usual way.

The next morning, he confirmed that he still wanted to withdraw. This was carried out in the following way:

He remained in bed as the NIV was still providing the best relief of dyspnoea and orthopnoea this was kept in place. Drugs were administered by the SC route:

All times in minutes

Time zero: 5mg each morphine, midazolam, levomepromazine

T60: in anticipation of reducing ventilator settings increased SC infusion morphine 20mg, midazolam 20mg and levomepromazine 25mg.

T90: Unsettled, 7.5mg each midazolam, levomepromazine 7.5mg.

Once settled progressive ventilator setting reduction every 10-15 minutes (back up rate and pressure). No further medication required.

T180: ventilator settings now IPAP 8cm and BUR 8bpm. 7.5mg each morphine, midazolam, levomepromazine in preparation for mask removal

T210: ventilator stopped and mask removed.

T375: some signs of distress, (frowning). 5mg each morphine, midazolam, levomepromazine.

T510: not looking as settled with increased fasciculations and some muscle spasms 7.5mg midazolam.

T 575 minutes: TD died peacefully.

5. After withdrawal

Documentation

All the professionals involved should make appropriate documentation of:

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

Following introduction of the Medical Examiner to England and Wales, the Medical Certificate of Cause of Death (MCCD) can be completed by a medical practitioner if they have attended the deceased 'in their lifetime', with no specified time constraints. The attending practitioner must then share the MCCD proposed cause of death with the Medical Examiner, who will scrutinise this before submission to the registrar. In Scotland the MCCD can be completed by a medical practitioner who has attended the deceased in 'their last illness'. The process for Northern Ireland stipulates that only the doctor that has attended the deceased in the last 28 days of their life can complete the MCCD.

The MCCD may read for example:

1a) Respiratory failure (due to) 1b) Motor Neurone Disease

For a patient that has withdrawn ventilation at home, a decision about who is best placed to complete the MCCD needs to be considered as part of the planning process

Standard 5

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

A care team debrief should be planned.

Support for the family

The family will need information and support to manage the immediate post-death processes (e.g. verification and certification of death, funeral director, 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 particularly at this time.

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

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

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

- Phone contact from a senior professional involved in the care of the patient in the first few days and a few weeks later.
- Signposting and phone numbers to who they could contact if they need support including MNDA connect: 0808 8026262 / mndconnect@mndassociation.org. MNDA Scotland MNDscotland.org.uk 0141 332 3903.

Support for paid carers and 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.

The working lives of some paid carers may have been dominated by the one to one care that they have given a ventilator dependent patient for many months or years. This should be recognised with some carers needing additional support from their employers at this time.

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.

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Appendices

Appendix 1. Resources to support professionals and patients and families

Motor Neurone Disease Association - http://www.mndassociation.org

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

Information Sheets 8B: *Ventilation in motor Neurone Disease (2022)* and 8C: Withdrawal of ventilation with MND (2021) have been included in this guidance in its entirety as a useful resource, and sign posts readers to other information that may be of value.

MND Scotland – www.MNDscotland.org.uk

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

Association for Palliative Medicine

The Association for Palliative Medicine of Great Britain and Ireland (APM) is an association for doctors and other professionals who work in or have a special interest in palliative care. The APM's Position Statement (2015) on the 'Withdrawal of ventilatory support at the request of an adult patient with advanced neuromuscular disease' is included in Appendix 8.

HMViP 2024 End of Life Care https://hmvip.co.uk/end-of-life

Hospice UK – https://www.hospiceuk.org/hospice-care-finder. Support and advice can be accessed via your local palliative care team. This website helps you identify your local palliative care services.

Media and online resources

Inside the Ethics Committee: *Withdrawing Treatment* (Series 7 Episode 3), **BBC Radio 4**. This podcast considers the issues that may be faced by patients, carers and healthcare professionals when deciding to withdraw ventilatory support in MND. http://www.bbc.co.uk/programmes/b012r7jn (accesses 8th November 2024)

Audio summary of findings of patient and families experiences by Dr Ellie Wilson, University of Nottingham on the science communication channel **SciPod** - Dr Eleanor Wilson | Breath by Breath: Decision-Making in the Final Stages of Motor Neurone Disease • scipod.global (Accessed 8th November 2024)

HealthTalk.org resource where patients and their families have shared their decision-making about and experiences of living with assisted ventilation by tracheostomy. https://healthtalk.org/introduction/tracheostomy-ventilation-and-motor-neurone-disease/

My Breathing – https://mybreathing.mymnd.org.uk/. A guide to using non-invasive ventilation for people living with motor neurone disease developed by Sheffield MNDA care team.

e-learning for health https://www.e-lfh.org.uk/programmes/end-of-life-care/. This Health Education England e-training resource for end-of-life care has sessions which support development of communication skills and advance care planning for health and social-care staff.

Resources for professionals and patients relating to advance care planning

Advance decisions to refuse treatment: A guide for health and social care professionals, 2013. Advance-Decisions-to-Refuse-Treatment-Guide.pdf (england.nhs.uk)

British Medical Association (2019) Best interests decision making for adults who lack capacity A toolkit for doctors working in England and Wales. *https://www.bma.org.uk/media/1850/bma-best-interests-toolkit-2019.pdf*

British Medical Association (2024) Ethics Toolkit: Mental Capacity Act in England and Wales. https://www.bma.org.uk/media/4z1l3khg/mental-capacity-act-england-and-wales.pdf

NHS Improving Quality (2014) *Capacity, Care Planning and Advance Care Planning in Life Limiting Illness: A Guide for Health and Social Care Staff* [Online]. ACP_Booklet_2014.pdf (england.nhs.uk)

MND Association: Information sheet 14A 2021.
Advance Decision to Refuse Treatment (ADRT) and advance care planning.
https://www.mndassociation.org/media/130#:~:text=Advance%20Decision%20to%20Refuse%20Treatment%20(ADRT)%3A%20You%20can%20write,new%20treatments%20to%20be%20introduced.
(INCLUDES A SAMPLE AND EXAMPLE ADRT FORM)

MND Association: End of Life Guide 2021 End of life: A guide for people with motor neurone disease End of life guide - standard PDF 2021 (mndassociation.org)

Ventilator guides

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

Appendix 2. Legal position and decision-making in practice

Whilst certain legal principles are generic and applicable across the UK, the law in relation to mental capacity differs in specifics between England and Wales combined, Scotland and Northern Ireland. However, the legal and ethical principles are the same under the devolved legislations. In Scotland the equivalent legislation is The Adults with Incapacity Act 2000, in Northern Ireland The Mental Capacity Act 2016.

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

1. Legal principles

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

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

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

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

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

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

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

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

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

This scenario, where the patient with capacity has decided that the burdens of continued medical treatment outweigh the benefits, is distinct therefore from where a patient with advanced neurological disease has chosen to foreshorten their life either actively with a method of suicide or under the umbrella of 'self-neglect' such as by deliberate sustained starvation.

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

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

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

2. Advance care planning

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

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

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

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

The patient has capacity.

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

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

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

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

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

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

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

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

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

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

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

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

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

You are required to maximise the person's capacity and to facilitate communication			
Question		**Y/N**	Answer
1	Does the person have the capacity either give consent or refuse treatment him or herself, with appropriate support where necessary		**YES**: The person has capacity to make the decision him or herself. The advance decision is not applicable. Ask 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 *all* 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 anticipated them?		**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. 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: • 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

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

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

If a patient is dying and lacks capacity, despite all measures to maximise capacity having been made, it may be appropriate to give consideration to discontinuing their non-invasive assisted ventilation on a 'best interests' basis'. This could be a clinical decision because of problems with the patient-ventilation synchronisation, secretion management or burdensome distress caused by some aspect of non-invasive assisted ventilation. In these cases, a formal best-interest decision may need to be made.

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

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

The doctor needs to evaluate:

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

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

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

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

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

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

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

The Mental Capacity Act (NI) 2016 (the Act) fuses together mental capacity and mental health law for those aged 16 years old and over in Northern Ireland within a single piece of legislation. The Act does not specifically legislate on advance decisions (as the Mental Capacity Act 2005 has done in England and Wales). The absence of a statutory basis for an advance decision means that the common law position will apply. The common law requires that to be valid an advance decision the person must have been aged 18 or over and been capacitous at the time it was made.

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

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

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

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

1. Commencing assisted ventilation

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

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

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

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

Future scenarios that are useful to touch on include:

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

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

Providing information about future scenarios and end-of-life choices whilst providing positive support to patients to get them through the initial hurdles is a challenge that will require both an advanced level of skill in communication and a team approach. Whilst it is unlikely to be appropriate to have a detailed discussion about withdrawal of NIV leading to death at this time, clearly there are some benefits in beginning such discussions at a stage of the illness before fatigue and communication difficulties increase.

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

The MNDA has written information available for patients and families (Leaflet 8b – Appendix 5).

Tracheostomy ventilation (TV) is not common in the UK but appears to be an option patients are increasingly aware of. It is becoming a positive choice made by some patients with MND. Discussion about TV use 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?

TV can be offered as a routine planned procedure particularly if significant bulbar symptoms make NIV less effective. The commonest scenario for elective 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. Resources are available on Healthtalk.org where patients and family talk about their experiences of using TV and decision-making related to this.

2. Established assisted ventilation

A patient may initiate conversation about their future with 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 health care professionals 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 may facilitate decision making 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 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 take in the future. The record of the discussion helps professionals understand how the decision has been shaped and how confident and settled the view of the patient is. In the specific context of a patient who may wish to withdraw assisted ventilation at a time in the future, it also allows the team to begin to develop an appropriate plan of who, how and where and to involve people at an early stage to support this plan.

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

The written record should include:

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

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

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

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

In almost all cases, the information should be kept with the patient. Most patients and families find this reassuring and helpful; only very occasionally would the patient or their family find this too distressing.

Appendix 4. 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. With the backup rate of inspiration provided by the ventilator the patient's chest may still move for a while 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.

Example care plan for care agency staff after a patient has died with NIV in place

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

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

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

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

Death verification: Guidance for verification of death (2024) includes the following instruction for the clinician in this circumstance: *It is recommended that the ventilator be switched off and continued checking for a pulse, alongside auscultating for the presence of a heartbeat, occurs. Following this, the [usual] verification process should be followed, ensuring all checks are conducted over the 5-minute period.* Care After Death guidance | Hospice UK

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

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

- Disconnect straps and remove face mask.
- Turn off the ventilator according to the instructions for that machine.
- Disposed of mask and tubing in general domestic waste.

Instructions for how to turn off the ventilator should be provided by the respiratory support team and in the patient's home

Contact the hospital or designated provider to arrange for collection of the ventilator when appropriate.

Appendix 5. Motor Neurone Disease Association information sheets

8B Ventilation for motor neurone disease (2022)

PDF

8B Ventilation for
motor neurone diseas

8C Withdrawal of ventilation with MND (2021)

PDF

8C Withdrawal of
ventilation with MND.|

Appendix 6. Glossary of Terms and Abbreviations

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

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

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

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

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

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

IPAP is inspiratory positive airways pressure, EPAP the expiratory positive airways pressure, BUR, the rate at which the ventilator delivers breaths which the patient does not trigger themselves.

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

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

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

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

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

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

Appendix 7. Contributors to Guidance development

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The help of Dr Gemma Andrew, Medical Adviser to the Coroner's Service of Northern Ireland, Fiona Wilcox, medico-legal secretary for the Coroners' Society of England and Wales, Dr Michael Devlin, Head of Professional Standards and Liaison the Medical Defence Union was also acknowledged.

Responses to the consultation in 2015

Responses were received from 24 doctors and nurses working palliative care, neurology, primary and ventilation care and the following organisations and clinical leaders.

National Clinical Director for Adult Neurology, NHS England	Dr David Bateman
Chair of the Royal College of General Practitioners ethics committee	Prof. Simon Gregory
Association of British Neurologists	Prof. Kevin Talbot
British Medical Association, Medical Ethics Committee	Veronica English
British Thoracic Society	Sally Welham
General Medical Council, Standards, Ethics and Education Policy	Mary Agnew
Hospice UK	Dr Ros Taylor
Intensive Care Society and Faculty of Intensive Care Medicine Joint Professional Standards Committee	Dr Gary Masterson
National Council for Palliative Care and Dying Matters	Claire Henry
Royal College of Physicians Joint Specialty Committees for Palliative Medicine & Clinical Neurosciences	Dr Simon Land
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Appendix 8. Association for Palliative Medicine Position Statement November 2015

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

This statement intends to set out the legal and ethical position for the care of patients with neuro-muscular conditions in the UK who request that their ventilatory support be withdrawn. While the ethical principles are generic and applicable across the UK, the law in relation to mental capacity differs between England and Wales combined, Northern Ireland and Scotland (see Appendix 1). For simplicity the rest of this document draws on the 2005 Mental Capacity Act (MCA) for England and Wales.

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

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

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

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

5. Whilst the timing of death will be influenced by the withdrawal of ventilation in these circumstances, the cause of death from a medical perspective remains the advanced neurological disease, and the classification of the death should be natural causes[‡‡] for the purposes of issuing a medical certificate of cause of death and subsequent registration of the death by the next of kin. Such a certificate may read for example:

   1a ventilatory failure (due to) 1b advanced motor neurone disease.

6. When a patient with capacity has decided that the burdens of continued medical treatment outweigh the benefits, this is distinct in law from a decision to foreshorten their life by suicide or 'self-neglect'.

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

8. Withdrawing a medical treatment from a patient who no longer has capacity, but who while having capacity made an advance decision to refuse treatment (ADRT) which is specific in this regard and valid for these particular circumstances, is not euthanasia even if the medical treatment is life-sustaining.

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

9. Withdrawing a medical treatment from a patient who no longer has capacity, on the advice or request from an appointed attorney, including decisions on life-sustaining medical treatment, and where on multidisciplinary review this request meets 'best interests' criteria, is not euthanasia even if the medical treatment is life-sustaining.

10. Withdrawing a medical treatment from a patient who no longer has capacity but who has not made an ADRT or appointed an attorney, is a conventional 'best interests' determination; the principles of which are set out within the Mental Capacity Act 2005 and refined within case law.

11. Patients and clinicians should openly discuss their thoughts and concerns about assisted ventilation and quality of life, and the circumstances in which a life sustained by ventilatory support would become intolerable or unacceptable. These discussions involving the patient, their family (with due regard for confidentiality) and the multidisciplinary team preferably should begin before assisted ventilation starts and continue throughout the duration of the illness.

12. Discussion of factors leading to the decision to stop assisted ventilation should be open, without coercion and thorough, seeking to identify any potential for alternative decisions and to minimise the impact of such a decision on family members. Ideally such discussion should be with the individual patient, family and healthcare team members, with these key people together.

13. Assessment of capacity to make the decision to stop ventilatory support is mandatory. As a matter of routine it should be a practitioner familiar with the issues who is assessing capacity for decision making on those issues. Given the challenges in such decisions, and in the enactment of Advance Decisions to Refuse Treatment, it may sometimes be advisable to involve more than one appropriately trained clinician in assessing the patient's capacity, and to gather feedback from the multi-professional team and the family regarding the consistency of the patient's wishes. Rarely this may require additional expertise such as that of a psychiatrist to determine whether there is an identifiable and treatable mental-health disorder compromising capacity.

14. The clinical conditions where ventilatory support is required to sustain life also involve conditions where patients often cannot physically withdraw assisted ventilation themselves and so it will need to be withdrawn by the clinical team.

15. Withdrawing assisted ventilation may lead to distressing symptoms that require anticipatory and timely treatment with appropriate doses of medications such as sedatives and opioids targeted at relieving these symptoms. As with all good practice in palliative care, the intent must be solely to avoid or ameliorate symptoms of discomfort or distress. Relieving a patient of discomfort and distress is a fundamental medical responsibility and is not a modifier of the cause of death as set out above.

16. This area of care is challenging and requires excellence in multidisciplinary working and clinical leadership. Input from specialist palliative care will be helpful and support for members of the team is important.

17. The GMC guidance *Treatment and Care towards the End of Life: Good Practice in Decision Making* (2010, updated 13.12.24) and *Decision Making and Consent* (2021, updated 13.12.24) 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.

The relevant law

1. 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:

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

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

4. *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')

5. 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

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

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

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