Pathway for Preparing to Withdraw Non-Invasive Ventilation (NIV) in Patients with MND

This is to be read in conjunction with the “Guidelines for Withdrawing Non-Invasive Ventilation (NIV) in Patients with MND”.

If patient is 24 hour dependent on NIV and decides to discontinue it, support and forward planning are essential.

Discuss with family end of life decision, what will happen and support needed.

**Identify keyworker to co-ordinate process**
- who prescribes and administers drugs (see ** below)
- route
- who removes mask
- who stops the ventilator

**CONSIDER SETTING**

HOME
- Family
- GP
- District Nurse
- Hospice @ Home
- Palliative Medicine Consultant
- MND CNS
- Carers
- Ventilation Nurse Specialist

HOSPICE
- Family
- GP
- District Nurse
- Palliative Medicine Consultant
- MND CNS
- Hospice Nurse
- Ventilation Nurse Specialist

HOSPITAL
- Family
- GP
- Palliative Medicine Consultant
- MND CNS
- Respiratory Consultant
- Ventilation Nurse Specialist
- Ward Staff

Ensure all members of MDT aware of patient’s decision and care plan and that any member of the team can withdraw from the patient’s care.

Ensure carers in place and identified doctor and nurse available for up to 48 hours.

Effective sedation sufficient to manage distress
- who will prescribe and administer and what route
- how the NIV is adjusted, by whom and who will remove the mask

Consider needs of family and professionals after death
- bereavement support for family
- debrief for professionals

**
Prescription of drugs is on an individual patient basis. See anticipatory prescribing in the “Guidelines for Withdrawing Non-Invasive Ventilation (NIV) from MND Patients”. Contact a palliative care doctor for guidance.
Leicestershire and Rutland MND Supportive and Palliative Care Group

Guidelines for Withdrawing Non-Invasive Ventilation (NIV) in Patients with MND

1 Withdrawing NIV in Patients with MND

Some patients may wish to discontinue their NIV as a matter of choice. It may be appropriate when discussing and/or initiating NIV to mention that it can also be stopped if they decide they no longer want it in the future.

It is unlikely to be appropriate to have a detailed discussion about withdrawal of NIV leading to death but clearly there are some benefits in communicating at an earlier stage of the illness before fatigue and communication difficulties may increase. While legally a competent patient is entitled to make such a decision, it may cause considerable concern for some members of the patient’s family or healthcare professionals and managers, as the patient’s choice may be viewed by some as tantamount to assisted suicide. However, what is taking place is the patient (or their proxy) taking an active decision to refuse a medical treatment or have it withdrawn, which from a medical ethics and legal perspective is not assisted suicide. Continuing with a treatment that the patient does not want is not legal and could be constituted as assault.

Complex emotional issues may arise around the withdrawal of NIV for families, carers and health professionals. These need to be addressed before the withdrawal of NIV, and the need for bereavement care should be addressed at an early opportunity. Staff support mechanisms, including the possibility of a staff de-briefing meeting, need to be considered. It is strongly recommended that the doctor leading the withdrawal discuss the situation with a doctor who has undertaken such care before. A list of such doctors should be available through the Association for Palliative Medicine.

The patient may choose to withdraw NIV at home or in an in-patient setting such as at the Hospice or Glenfield Hospital.

2 Background Information about Ventilation

Normally patients with MND will be on Pressure Control Mode. In withdrawing NIV the mode does not need to be altered. However, the Inspiratory Positive Airway Pressure (IPAP) can be reduced by 50% to ensure the patient is sufficiently sedated to undertake complete withdrawal of NIV.

Familiarisation with the ventilator is crucial before ventilation withdrawal takes place. It is important to know how to:

- change the pressure settings
- turn off or adjust alarm settings
- turn off the machine

Generally, the preferred method for discontinuing ventilation would be to provide adequate anticipatory sedation for the patient, testing this by reducing the pressure level on the NIV to 50% for a few minutes and seeing if this causes any distress. Sedation can then be increased if needed. This would be followed by a trial of mask removal and checking the
patient is not distressed before then switching the NIV machine off. It is key to ensure maximum palliation of dyspnoea and other symptoms before stopping NIV.

3 **The Process of Withdrawing NIV**

When a patient using NIV requests its withdrawal, the reasons for the request should be explored and the other options available considered. Given all the issues around the Liverpool Care pathway and the potential of relatives’ interpretations, there should be discussion with patient and family on two separate occasions and ideally with two different senior health care professionals.

The family must also have an opportunity to ask questions and express concerns.

It is important to be clear that if a patient dependent on NIV 24 hours a day stops it, they are likely to die shortly afterwards – from a matter of minutes, or sometimes extending to a few hours or even days.

**If a patient is not 24 hour NIV dependent**, they may choose not to continue with NIV at any point by simply not replacing their mask. This may lead to increased dyspnoea requiring appropriate symptom control. Consider starting a syringe driver the day before the mask is removed if there are uncontrolled symptoms of anxiety or breathlessness.

**If a patient is 24 hour NIV dependent** and wishes to discontinue its use, the principles are the same but death is likely to follow more closely. Forward planning is required to ensure that the patient does not experience distressing symptoms at the time of withdrawal.

4 **Involvement of the MDT**

Discuss with and inform the wider team of the patient's decision (MND CNSs, GP, district nurse, Hospice @ Home, SLT, dietician, OTs, physios, complementary therapists).

Ask team members' views and give anyone who does not wish to be involved the option of withdrawing from that patient's care.

Which professionals need to be present will vary from case to case. This should be planned ahead to ensure availability. There will need to be a nurse and doctor available, possibly for up to 48 hours. This will usually be members of the specialist palliative care team.

5 **Anticipatory Prescribing**

There will be variation in how much medication is required for different patients though a combination of benzodiazepine and opioid is usually recommended. For patients completely dependent on NIV, they are likely to require deep sedation to unconsciousness to tolerate its withdrawal.

Our experience has been to start with 10mg morphine and 10mg midazolam subcutaneously and assess after 20 to 30 minutes. Alternatively medication may be given intravenously in aliquots of 5mg. The intravenous route allows more rapid and flexible titration to symptom management and may reduce the time the process of withdrawal takes.

The total dose may range from 20mg of morphine and midazolam to higher doses to achieve the adequate depth of unconsciousness to manage any respiratory distress. Discussion, including input from a palliative medicine consultant prior to the event is advised.
A syringe driver should be considered if death may not occur rapidly such as when a patient is not 24 hour NIV dependent.

Levomepromazine may be a useful 2nd line sedative, especially if a patient is benzodiazepine tolerant - suggested initial dose of 25mg stat.

6. Practicalities

**Discuss with the patient and family:**

(i) Who wants to be there at the time of death?
(ii) Who will remove the mask - professional or family member?
(iii) The possibility of gasping, changing colour, etc.
(iv) The uncertainty in how long it may take to die after stopping NIV. Try and make the expectations realistic.

**Differences for patients at home** compared to hospice inpatients need to be considered, such as the availability of professionals who may need to be present for several hours in a patient's home, and the provision of privacy at home when professionals are also present. The GP is much more likely to be involved with a death at home.

**After death**
Bereavement support should be available to the family and carers acknowledging the complex emotional impact of withdrawing NIV. Debriefing and support should be available to the professionals involved.

**Acknowledgements**
Based on a Clinical Guideline from St Wilfrid’s Hospice, Chichester written by: Brendan Amesbury and Kathy White

These Guidelines have been written by a working party consisting of:
Alison Conway (Respiratory Clinical Nurse Specialist), Jo Smith (Respiratory Specialist Physiotherapist) Dr Luke Feathers, Dr Christina Faul, Dr Barbara Powell & Dr Dave Riley (Consultants in Palliative Medicine), Jane Glover & Jo Joyce (MND Clinical Nurse Specialist), Benny Rossi (LOROS Day Therapy and Community Services manager), Jane Skelton (MND Association), Rachel Boothman (MND Association), Kris Albrow (District Nursing Sister), April Andrews (Staff Nurse, Hospice at Home), Mo Fisher (Staff Nurse, Hospice at Home), Fiona Brant (Staff Nurse, Hospice at Home).

Thanks to the following who gave useful advice about this guideline: Nigel Sykes; Mary-Ann Ampong; David Oliver.

**References**

GMC End of Life Care Guidance 2013
King's College Hospital NIV in MND guidelines.
Report from West Sussex Dilemmas and Good Practice study day, 2006.


Association of Palliative Medicine position statement on ventilation withdrawal