



MND Podcast number three:
Respiration and Non-invasive ventilation

Hello my name is Robyn Reid. I am the MND Shared Care Worker in the Southern Region of Melbourne. Welcome. This is a series of podcasts on Motor Neurone disease. This podcast may help you to understand the issues around respiratory function in people with MND and Non-invasive ventilation. The person with MND will have symptoms that get worse over time and this includes their ability to breathe and get enough oxygen. This is called Dyspnoea. This is caused by weakened respiratory muscles- the intercostals, diaphragm, and abdominal muscles. The lack of air in the lungs can reduce the volume of speech as well.

As respiratory function slowly worsens the person may wake in the mornings with headaches, feeling “foggy” and not alert, increasingly short of breath and sleepy during the day and having sleep disturbances. These are the symptoms of hypoventilation. Breathing is more difficult in sleep due to decreased muscle tone, including the diaphragm, decreased breath volume, and decreased ventilation. Lying flat at night can make dyspnoea worse. This means there is less oxygen and more carbon dioxide circulation, and as a protective mechanism the brain wakes the person to increase breathing. This means broken sleep. The respiratory drive in the brain stem becomes affected. This gradually affects daytime ventilation as well.

The person with MND will then undergo assessment through sleep studies and respiratory function tests with the Victorian Respiratory Support Service and if appropriate will be offered Non-invasive Ventilation (NIV). Not all people with MND will have Non-invasive Ventilation as it can prolong life, but it does



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improve quality of life, and enables the person to feel more alert.

Therefore, they are more likely to have energy, and wish to participate in activities and socialise with their loved ones. This is an important consideration for someone with a life limiting disease. Initially the person will only use the NIV at night, but gradually people become dependent on it for well-being most of the time. The benefits of NIV at night mean less changes in oxygen and carbon dioxide levels so that they are normalised. It stabilises ventilation, improving sleep and returning the respiratory drive to normal.

Once the assessments are complete and the person has agreed to NIV then they will be given a prescription of settings specifically for them. Breaths are set within a window and allow for voluntary breaths as well. The machine is not a CPAP machine which has constant air pressure to keep the airway open. CPAP machines are commonly used for snoring and sleep apnoea. This is a VPAP or Variable Positive Air Pressure, or BIPAP, Bilevel Positive Airway Pressure which takes into account breathing in and out. On inhalation the machine increases the pressure and on exhalation the machine decreases the pressure according to the settings prescribed for that person. These are locked into the machine and cannot be altered without a code. Giving extra oxygen is not recommended or indicated for people with MND. Not everyone can tolerate NIV. It is difficult to allow something to cover your face when you are struggling to breathe.

The air from the machine is quite forceful and to add to the comfort of the person using the equipment, the machine has two measures. The first is a



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humidifier attached to the machine to put moisture into the air that stops the respiratory system from drying out. The second is a ramp button so that the air flow can be adjusted to the person's needs and increase air volume slowly. If you are having difficulty breathing a sudden gust of air into your lungs would be uncomfortable, so the ramp button allows the person control over the volume of air when turning on the machine. The humidifiers need to be removed when moving the person and their machine to prevent backflow into the unit. The water in the humidifier can be ordinary tap water but it needs to be replaced daily. A weekly clean with soapy water is all that is needed. Filters at the back of the unit must be replaced every 2-3 months.

People who are dependent on NIV will have a spare machine supplied by the Victorian Respiratory Support Service, who continues to monitor and assess the person and their respiratory function regularly. They provide a 24 hour service across the state of Victoria and offer an outreach service. Their contact details are able to be downloaded in the document of this podcast. If the person has concerns about their equipment make sure they bring the equipment with them to their review. The NIV is portable and runs on battery which is able to be recharged and lasts several hours.

There are face masks and nasal masks of varied shapes and sizes to get the best fit for the person. It is critical that there are no leaks around the mask. Apply the mask squarely to a clean face and with equal tension on the headgear, firmly, but not tightly. Check that it is not too tight on the bridge of the nose as this can cause a pressure area. Some small leaks around the top of



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the mask near the bridge of the nose are a trade-off to prevent pressure sores. As muscle weakness makes it difficult to take off the mask, make sure there is a buzzer or bell that is appropriate. Headgear may be modified to allow the person to remove it easily. This is important during a coughing fit, or to attend to secretions, or while talking or eating. This can add to the carer's burden of care if they need to constantly take the mask off and on around the clock when the person with MND can't remove it. Wipe the mask cushion with a wet cloth and wash mask and headgear in warm soapy water, rinse and allow to dry. Mask cushions have a life of approximately 18 months. Masks are constantly being changed and monitored for correct seal due to muscle deterioration, weight loss and changing needs.

Sometimes the humidification will cause noisy rattling to build up in the tubing caused by condensation and room temperatures. This can be managed by turning down the humidification, warming the hoses by putting them under the bedclothes, or wrapping them to keep them warm, or heating the room. Skin may become broken when masks are constantly being taken on and off or from pressure. Contact VRSS if this is happening. Protective dressings are available such as Mepilex lite or Aquaclear.

Care when using NIV is important in the following areas. Bowel care is essential as a full abdomen can stop the diaphragm from functioning properly. The build-up of air can be vented through the Peg tube or using the ramp function on the machine to stop air being put into the stomach. Small regular meals are best and adequate posture and support of the neck while eating to help



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prevent aspiration of food and fluids. People may choose to use a nasal mask while eating.

Being short of breath creates anxiety, and anxiety further increases shortness of breath. Controlling anxiety increases well-being, and can reduce the settings on the NIV. Anxiety may mean prescribing appropriate medication.

Breathlessness can be assisted with small doses of morphine. When showering move the NIV into the bathroom in case it is needed and make sure it will not get wet. Shower early in the day to reduce the impact of fatigue. Have a fan for air flow if that helps to relax the person. Have the persons back to the water to reduce breathlessness in the shower.

If secretions are an issue then NIV may be difficult if the mask is being removed frequently. Seek a review and management of secretions. Some people use a machine called an Insufflator/Exsufflator that assists to remove secretions from the lungs by helping to increase the strength of coughing by providing a vacuum.

If the person has a coughing fit remain calm, open a window or use a fan to increase circulating air. Provide calm reassurance until the coughing fit passes. The person may be afraid they will choke to death but they should be reassured that death caused by choking attacks is rare and that the final stages of MND are usually peaceful and dignified.



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Treatments aimed at improving respiratory function in MND include: Assisted cough technique, physiotherapy, flu vaccination, early management of respiratory infections, secretion management, adequate nutrition and hydration, ventilation, and attention to posture and muscle supports. Complications of NIV can include: Aspiration of secretions, air in the stomach, pressure areas from masks and straps, air leaks making ventilation ineffective, and nasal symptoms such as dryness, congestion, runny nose, or nose bleeds.

In Australia people with MND are not treated with tracheostomy as this would result in being “locked in” – a situation where the person is totally dependent on ventilation and only being able to move their eyes as all voluntary muscles are not working, and would be completely dependent on artificial ventilation indefinitely. People with NIV even if they are getting assistance to breathe with via a mask and machine, will die of respiratory failure.

MND is a unique disease in that people may decide that the advantages of NIV are no longer contributing to their quality of life and they may decide to withdraw the ventilation. This will mean that their disease and their current respiratory function may not be able to keep them alive without it. This means they may choose to die. This discussion around choices and options happens when the equipment is supplied and is an ongoing one. This should be a planned event with the appropriate team and medications available for the person. Should they request withdrawal of ventilation, medication and



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intensive support via the VRSS team and the palliative care team will be provided if they are in their own home and in hospital or hospice settings.

It is of utmost importance that VRSS have the respiratory function tests completed that support the withdrawal of Non Invasive Ventilation. This means that when the NIV is withdrawn the person's respiratory function is such that death will be happen quickly following withdrawal. The worst case scenario is that NIV is withdrawn and death does not occur for some time causing increased distress to those participating. Consultation and communication are very important in this process to ensure the roles and responsibilities are clear and that the necessary supports and medications are in place in the planned event of NIV withdrawal.

It is extremely important that conversations about end of life planning happen early in the course of the disease and the person's wishes about certain treatment decisions are known and documented and communicated to all caregivers. This is urgent due to the possibility of the impact on communication of poor speech or no speech, and the possibility of cognitive impact around decision making and understanding. In a small number of people with MND they may go on to have frontotemporal dementia which may reduce their capacity for making decisions about their treatments and end of life wishes. Advance care planning, Refusal of treatment certificate and Medical Treatment Decision Maker documentation should be noted and made available when necessary. Loved ones may find the decision to withdraw ventilation distressing and may need extra support to understand the process.



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Professionals may also find the process difficult unless they understand they are withdrawing an unwanted treatment and allowing the disease to take its natural course. There are protocols for providing the withdrawal of ventilation which you can find on the document accompanying this podcast.

I look forward to discussing more detail about various aspects of MND management in future sessions which will be available on the SMPPC website. I would like to take this opportunity to give my thanks to staff at Bethlehem Calvary Health care and MND Victoria and VRSS for the support and training received in my role as MND SCW in the Southern Region. Please feel free to contact me should you wish to have an education session on MND for your agency or to receive a secondary consultation. You can contact me on: 0428264446 or email me at: Robyn.reid@smrpcc.org.au

References:

1. VRSS non-invasive ventilation-practical tips for carers and health professionals. Presentation MNDV
2. Power point presentation Withdrawal of Non Invasive Ventilation. How can we help all involved? Dr David Oliver University of Kent
3. Non Invasive Ventilation 2008 Dr Mark Howard, Director VRSS, Austin Health
4. Withdrawal of ventilation at the patient's request in MND: a retrospective exploration of the ethical and legal issues that have arisen for doctors in the UK
Phelps K, et al. BMJ Supportive & Palliative Care 2015;0:1–8. doi:10.1136/bmjspcare-2014-000826
5. A Problem solving approach for General Practitioners and the Primary health team 2000 3rd Edition The MND Association
6. VRSS Ward 5 Austin Hospital Outreach service Phone: 94963665
After hours Ward 5 west Phone: 94963685
7. Example of protocol for withdrawal of NIV

Title: Withdrawal of Non-Invasive Ventilation (NIV)

Section: Clinical Practice PROTOCOL

Person Manager Clinical Services & LC and Manager of Client Support Services

Responsible:

Approved by: Policy & Procedure Committee **PURPOSE**

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Clients admitted to Palliative Care Service (PCS) wishing to withdraw their NIV must be referred to a specialist service i.e. Victorian Respiratory Support Service Austin Health (VRSS AH) who will be responsible for planning and managing the withdrawal.

Note the procedure cannot be performed unless the client is completely physically dependent on NIV. A medical doctor or a VRSS outreach nurse in liaison with a respiratory physician will be responsible for determining if a client is physically dependant on NIV.

PCS will not lead the withdrawal of NIV under any circumstances.

This policy will apply to clients who are using NIV and are conscious (awake), semiconscious, or unconscious.

It will apply to clients when one or more of the following factors are present:

1. The client is experiencing an illness in which ventilation is postponing an inevitable death, and prolonging suffering.
2. The benefits of continued ventilation do not medically justify the burdens imposed on the client.
3. Client or MEPOA (Medical Enduring Power of Attorney)/ guardian wishes ventilation to be withdrawn.
4. The clients' best anticipated outcome from continued ventilation is a state of health inconsistent with their wishes as documented in the medical record, advanced care plan, indicated by MEPOA or as understood by their family.

Ethical considerations

Complete withdrawal of Non-Invasive Ventilation (NIV) will allow death to occur from the client's underlying disease process. Competent clients have the right to refuse any unwanted treatments, or request these treatments to be ceased (Victorian Medical Treatment Act 1988).

Withdrawal of ventilation will result in the client's death, but is not euthanasia. It is the underlying medical condition that is the cause of death, and the removal of NIV is simply the cessation of an unwanted or inappropriate treatment

DEFINITIONS

Withdrawal of Non-Invasive Ventilation (NIV): is withdrawal of non-invasive ventilation (i.e. BiPAP and VPAP), from the conscious (awake), semiconscious or unconscious person, allowing death to occur from the patient's underlying disease process (Ibrahim, 2012).

BiPAP and VPAP: is a form of positive pressure ventilation. BiPAP and VPAP machines have two phases of operation for each breath. IPAP (Inspiratory Positive Airway Pressure) provides a large breath and EPAP (Expiratory PAP) helps improves gas exchange and maintains the airway if there is any OSA. BiPAP and VPAP machines are used by people who need mechanical support to maintain normal ventilation. The machine will respond to the user's breathing efforts with no upper limit on rate, or will deliver breaths at the set rate (maximum 30) should he/she fail to take a breath (Personal communication, 10th of February, 2015, Anne Duncan).

PROCEDURE

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Once a client has indicated that they want to withdraw their NIV a case conference will be held with the GP, both clinical managers and other relevant staff to ensure the correct planning is put in place and a referral is made to VRSS AH to oversee the withdrawal of NIV.

The VRSS AH Outreach Service can be contacted on telephone number: (03) 9496 3665 during business hours Monday to Friday.

If the client is also known to Calvary Health Care Bethlehem CHCB, the clinic team can be contacted on telephone number: (03) 95953355 during business hours Monday to Friday.

All staff and volunteers at Palliative Care Service will respect clients' wishes and not impose their own values or beliefs on a client's decision to withdraw NIV.

The Manager of Clinical Services and Learning Centre and/or the Manager of the CSS team must refer any clients wishing to withdraw their NIV to a specialist service i.e. VRSS AH who will be responsible for arranging and facilitating the NIV withdrawal procedure.

A case conference will be held with VRSS AH (and CHCB if involved) and the Manager of Clinical Services and Learning Centre and/or the Manager of the CSS team to decide PCS's role in the withdrawal of NIV procedure. Nursing and/or CSS staff at PCS may be involved as a secondary support service to support clients and their families during NIV withdrawal but under no circumstances is PCS to lead or initiate a NIV withdrawal procedure.

The Manager of Clinical Services and Learning Centre and/or the Manager of the CSS team will decide which staff members will be present (if any) for the withdrawal of NIV procedure based on the staff members experience and beliefs and clients' needs.

The decision to withdraw ventilation treatment should be clearly documented by Nursing and CSS staff at PCS in the client record under the 'Advanced Care Planning/End of Life Care' section of client records.

A planned day and time to start withdrawal needs to be agreed on by the appropriate care provider(s) i.e. staff from VRSS AH in consultation with the client and their family/significant others. The client's General Practitioner (GP) and/or relevant medical specialist(s) and health care professionals e.g. CHCB, MND regional advisor, and staff at PCS need to be informed of the plan for withdrawal of NIV. The NIV withdrawal procedure cannot always be accommodated in the community and in such cases the client must be admitted to an inpatient facility.

A NIV withdrawal procedure in the community can only start between Monday to Thursday 1000- 1300hrs (excluding public holidays).

The people performing the NIV withdrawal procedure in the community will provide advice and support to PCS should such a situation arise.

Within 72hrs post the NIV withdrawal procedure, a debrief session will be held at PCS involving all staff involved.

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Adapted from: Ibrahim, M. (2012). Austin Health - Withdrawal of Ventilation in the Victorian Respiratory Support Service – Guidelines & Clinical Procedure.

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RELATED POLICIES

- Advanced Care Planning

REFERENCES

- Ibrahim, M. (2012). Austin Health - Withdrawal of Ventilation in the Victorian Respiratory Support Service – Guidelines & Clinical Procedure.

LEGALISATION

- Victorian Medical Treatment Act 1988.

Document reviewed and approved by Victorian Respiratory Support Service Austin Health (VRSS AH) and Calvary Health Care Bethlehem (CHCB) 17.04.15