

MND symptom management at end of life

Comprehensive symptom control and optimal psychosocial support is essential in the management of a peaceful and dignified death.

Increased involvement by a palliative care physician/team at this time is integral to optimal symptom management towards the end of life.

MND Australia 2014

The terminal stage is recognised as progressive weakness and often a sudden deterioration in a few days or hours.

The most common cause of death is respiratory failure, usually following upper respiratory tract infection.

- the terminal phase may be preceded by reduced chest expansion, a quietening of the breath sounds, use of accessory muscles (if any are left) and morning headache from CO₂ retention overnight
- signs may be noticed by carer or a member of the multidisciplinary team.
- the family can be prepared for the coming days and the patient's imminent death (after many months/years of disease)
- this may help prevent the shock of an 'unexpected' death or the family inadvertently ringing an ambulance after the patient has died at home.

Note: It is important to reassure patients and carers that death from choking is rare.

Check all symptom control and support needs

For management guidelines refer to other sections in this website related to:

- pain
- dyspnoea
- dysphagia, dry mouth and excessive and/or tenacious salivation
- equipment
- dysarthria
- psychological support

Reassess the needs of the carer and family and liaise with palliative care team.

Miller and others 2009b

What treatments reduce pain and dyspnoea in the terminal phase of ALS?

- Pain and dyspnoea are common correlates with suffering (Class IV) and a desire for physician-assisted suicide in late-stage ALS (Class IV).
- *Conclusion*
 - No controlled studies examined treating pain or dyspnoea in late-stage AL

- *Recommendation*
 - There are insufficient data to support or refute specific treatments for pain and dyspnoea in late-stage ALS (Level U)

What is the optimal method of withdrawing both noninvasive and invasive ventilation in ALS?

- Case series offer practical advice for withdrawing both invasive and noninvasive ventilation from patients with ALS (Class IV).
- *Conclusion*
 - There are no controlled studies examining withdrawal of ventilation in ALS.
- *Recommendation*
 - There are insufficient data to support or refute specific strategies for withdrawal of ventilation in ALS (Level U).
- Clinical context
 - Protocols based on consensus for withdrawal of mechanical ventilation in intensive care units (Class IV) include counseling and symptom control with opioids, benzodiazepines, and anticholinergic medications. We could find no controlled studies in any disease.

Mitsumoto and others 2005

Symptom management

Table IV addresses medical management during the last days and last hours of life. Advance directives may play an important role in directing symptom management, especially as pertaining to the last hours of life.

TABLE IV. Recommendations for symptom management during last hours of life

Terminal management of patients on ventilatory support

Recommendations to the field for development

1. Routinely assess patient's preferences for ventilatory support at successive stages of illness.
 - Provide assurance that respiratory distress will be actively managed with or without ventilatory support.
2. Discuss when to withdraw ventilatory support and what to expect at this time. Review advance directives with patients and family members.
3. Establish the basis for withdrawal of ventilation prior to initiating ventilation. Discussion should include:
 - The expected manner and time course of death
 - Medications that will be used to manage symptoms
 - Possible use of sedation
4. Withdraw ventilation with a physician present. Make all arrangements prior to the removal of support including discussing, planning, and implementing all cultural or religious rituals
5. Use parenteral medications, such as opioids and benzodiazepam, to achieve rapid sedation without paralyzing agents. Once comfort has been obtained, discontinue positive expiratory pressure, followed by conversion to a T-

piece in the case of tracheotomy ventilation.

Management of patients not on ventilatory support

Recommendations to the field for development

1. Follow the same procedures stated above except initiate medications (such as opioids) in a more gradual fashion (around the clock if distress recurs) since there is no specific event to anticipate.
 - Ensure that the physician is readily accessible for adjustments of medication.
 - Stop use of all unnecessary medications.
 - Consider discontinuing monitoring (vital signs, oximetry).

Research and program development recommendations

1. Develop treatment and management protocols/algorithms for alleviation of respiratory symptoms through the end of life.
2. Develop treatment and management protocols/algorithms on how to withdraw ventilatory support at end of life.
3. Undertake studies to examine how patients die in a natural setting as compared to those patients on ventilatory support.