ALS Palliative Care

Christen Shoesmith, MD FRCPC
Medical Director of the Motor Neuron Disease Clinic at LHSC
October 22, 2018
Disclosures

* I have no potential conflicts of interest to disclose
Objectives

1. Discuss palliative care for ALS patients at end of life
2. Discuss NIV discontinuation in patients who are fully ventilated
3. Discuss potential for organ donation from in ALS
4. Discuss MAID conversations in ALS
Palliative Care Guidelines

• 2008 AAN Practice Parameters
  ◦ Insufficient evidence was available to make useable palliative guideline statements

• Canadian Guidelines are being developed
1. Whenever possible, offer input from a palliative care team early in the course of the disease.

2. Initiate discussions on end-of-life decisions when the patient asks or provides an opportunity for discussion on the provision of end-of-life information and/or interventions.

3. Discuss the options for respiratory support and end-of-life issues if the patient has dyspnoea, other symptoms of hypoventilation or a forced vital capacity below 50%.

4. Inform the patient of the legal situation regarding advance directives and the naming of a healthcare proxy. Offer assistance in formulating an advance directive (GCPP).

5. Re-discuss the patients preferences for life-sustaining treatments every 6 months (GCPP).
6. Initiate early referral to hospice or homecare teams well in advance of the terminal phase of ALS (GCPP).

7. Be aware of the importance of spiritual issues for the quality of life and treatment choices. Establish a liaison with local pastoral care workers to be able to address the needs of the patient and relatives (GCPP).

8. For the symptomatic treatment of dyspnoea and/or intractable pain, use opioids alone or in combination with benzodiazepines if anxiety is present. Titrating the dosages against the clinical symptoms will rarely if ever result in life-threatening respiratory depression (GCPP).

9. Terminal restlessness and confusion because of hypercapnia can be treated with neuroleptics (e.g. chlorpromazine 12.5 mg every 4–12 h po, iv, or pr) (GCPP).

10. Use oxygen only if symptomatic hypoxia is present (GCPP).
1.7.1 Offer the person with MND the opportunity to discuss their preferences and concerns about care at the end of life at trigger points such as: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity.

1.7.2 Be prepared to discuss end of life issues whenever people wish to do so.

1.7.3 Provide support and advice on advance care planning for end of life. Topics to discuss may include:

- What could happen at the end of life, for example, how death may occur.
- Providing anticipatory medicines in the home.
- Advance care planning, including Advance Decisions to Refuse Treatment (ADRT) and Do Not Attempt Resuscitation (DNACPR) orders, and Lasting Power of Attorney.
- How to ensure advance care plans will be available when needed, for example, including the information on the person's Summary Care Record.
- When to involve specialist palliative care.
- Areas that people might wish to plan for, such as:
  - what they want to happen (for example, their preferred place of death)
  - what they do not want to happen (for example, being admitted to hospital)
  - who will represent their decisions, if necessary
  - what should happen if they develop an intercurrent illness.
Palliative Care Referrals in ALS

- Potential triggers for referral to a palliative care physician or team:
  - Patient request
  - Initiation of BIPAP
  - Significant respiratory insufficiency
  - Feeding tube insertion or refusal of tube
  - Prediction of death in less than 3-6 months
  - Individuals who become home bound
Death in ALS

- Approximately half of patients in US die at home or hospice facility, 25% of patients die in a hospital, 20% in a nursing home. Goutman, ALS. 2014 Sep;15:440-3.

- Death usually slow and predictable and less common to be sudden (6%, likely due to mucous plug or PE) or in sleep (6%). Acta Neurol Scand. 2010 Sep;122(3):217-23.
  - Education about these types of death may be useful
    - Especially indicating that death by choking is extremely rare
  - Majority die from slow respiratory failure
    - Initially become somnolent from hypercapnia
    - Often needing only small doses of narcotics for respiratory comfort (due to hypercapnia)
    - Very rare exceptions of non-ideal deaths
End of Life Care

- **Air hunger:**
  - Managed by
    - Oxygen
    - Morphine/dilaudid
    - Benzodiazepines
    - Fan/open window

- **Anxiety - benzodiazepams**
- **Pain – morphine/hydromorphone/fentanyl**
- **Oral secretions - glycopyrroloate**
Case 1

- A patient has advanced ALS with respiratory insufficiency and quadriplegia. He is dependent on his spouse for all activities. At his clinic visit, he complains of difficult to clear bronchial secretions which he cannot cough up.
  - What are his options?
Pulmonary secretions

- Often challenging to treat
- Ensure hydration status adequate
- Oral suction +/- deep suction
- Manual cough assist +/- breath stacking
- Mechanical cough assist
- Can try:
  - Oral therapies for sialorrhea, but often makes secretions thicker and more difficult to clear
  - Propranolol 20mg bid
  - Nebulized N-acetylcysteine (3-5 mL of 20% solution tid)
  - Trial of eliminating dairy
  - Sugar free citrus lozenges, grape seed oil, papaya enzymes
- Rarely tracheostomy is required for pulmonary toilet
Cough Assist

- Manual:

- Mechanical

Cough assist machines now provided by VEP pool
Case 2

- A 67 year old patient with ALS is on BIPAP for most of the day. He can only tolerate being off BIPAP for 2 minutes at a time. He is still ambulatory, but has severe bilateral arm weakness. He finds life intolerable and wishes to die. How can he be supported in death?
NIV withdrawal considerations

- Discontinuing NIV is like discontinuing a ventilator
  - Need to anticipate sudden and prominent dyspnea symptoms, particularly for those on >22hr/day
  - Should consider emotional reactions of staff who will have difficulty understanding withdrawal appropriateness (some may equate with euthanasia/MAID)
NIV withdrawal procedures

- Leicestershire and Rutland MND Supportive and Palliative Care Group
  - Guidelines for Withdrawing Non-Invasive Ventilation (NIV) in Patients with MND
- St Wilfrid’s Hospice, Chichester
  - Withdrawing Non-Invasive Ventilation from MND patients
Process

Discussion with patient, family/carers and involved professionals is required to ascertain that they all understand that:

1. A competent patient has the right to stop any treatment when they wish.
2. Stopping NIV will result in death in minutes, hours to days.
3. Complying with the patient's wishes is good medical practice not physician assisted suicide

Where should NIV withdrawal occur? Home? Hospice? Hospital?
LHSC NIV withdrawal process

Check list for Day Prior to Discontinuation

1. Nurse who will be caring for the patient on the day of discontinuation is assigned the patient.
2. A physician with experience prescribing palliative care medications meets with patient and family.
3. Social work is consulted to meet with the patient and family.
4. Medication route and types are planned.
5. Check with the patient what non-pharmacologic care they would find comforting (mouth care, bath, shower, blanket, etc.)
6. Spiritual care meets with family if desired.
7. NIV settings remain the same.
8. Celebration of life occurs in patient’s room (if desired by patient).
Day of Discontinuation of NIV

1. A **general floor staff briefing** is held to explain the procedure and appropriateness.
2. The nurse should be assigned **1:1 care** for this patient with regular breaks.
3. NP or physician to be present in the room for the first two hours after discontinuation of NIV.
4. Opioid bolus given +/- benzodiazepine given and titrated until the patient is comfortable and not in distress. A basal rate of the opioid is then infused, and the rate can titrated.
5. Once patient is comfortable, RT turns off the begins **wean of NIV** at 5min intervals via rate reduction first, then wean pressure support next (to IPAP of 8 cmH20 and EPAP of 4 cmH20) and finally removes the device completely. As each step of the wean, medications would be titrated appropriately.
6. Medication **boluses given q5min PRN** for any signs of patient distress. Opioids and benzodiazepines can be considered. Frequent boluses may be required for patient comfort
7. Once the patient has died, **debriefing** with the bedside nurse should occur to reflect on the experience and recommendations. There should also be debriefing for all unit staff so that the information spread throughout the unit is accurate and supportive.
Case 3

• A 57 year old man had a 6 month history of progressive extremity weakness which he attributed to worsening arthritis.
• Exam findings and investigations were consistent with ALS.
• He had increasing difficulties with orthopnea.
• Was initiated on BIPAP and use increased such that he was using it for most of the day.
Several months down the road, he decides that he wishes to die. He asks if he could be an organ donor. Would organ donation be an option?
Key Questions

- Is ALS transmissible?
- Should ALS organs be used for transplantation?
DCD Organ Donation

- Donation after Cardiopulmonary Death (DCD)
  - Can occur if:
    - Patient is ventilator dependent
    - There is no hope of reasonable recovery
    - Patient and/or family consents to donation
  - Organ assessment and transplant suitability occur prior to withdrawal
  - Life support is withdrawn and palliative medications are administered.
  - After the heart stops beating and there is no pulse for 5 minutes, death is pronounced.
  - The body is then taken to the OR for immediate organ recovery.
- For ALS patients to donate organs, they must either be predicted to die within 90 minutes of discontinuation of life support (BIPAP or invasive ventilation) or elect to have MAID
Scientific arguments against transmissibility of ALS

- Rates of conjugal ALS are very low (only 20 cases reported in literature, Dewitt et al 2012)
- Only one published case report of ALS developing in a solid organ transplant patient (liver transplant, Broering et al 2004)
- No animal to animal transmission of ALS reported
- The Blood Brain Barrier may protect against transmission through solid organs
Scientific arguments for transmissibility of ALS

• Prion hypothesis:
  ◦ Both SOD1 and TDP-43 exhibit the potential for seeded aggregation in vitro (Furukawa et al, 2011, Munch et al 2011)
  ◦ Cashman and Pokrishevsky’s evidence that misfolded wild type SOD1 can spread between cells in vitro

• Three cases of early onset ALS in recipients of human growth hormone from cadaveric pituitary extract, with ALS developing at least 10 years after use of GH.
  ◦ Does not confirm causation, but indicates transmission would take at least 10 years
Trillium Gift of Life Network

- Supports donation of organs from patients with ALS as an exceptional donor
- Supports donation into patients as a life saving need
  - Corneas and skin not accepted
  - Large organs accepted
Back to the case

- After much discussion, organ donation is presented to the patient and how it would occur. He agrees to pursue this option.
- Much discussion at the hospital about timing/availability of staff and OR
- DCD procedures followed in the anteroom to the OR on a Saturday with family in room.
- BIPAP is turned off and he dies quite quickly.
  - Unfortunately, liver lesion seen on OR organ recovery and no organs donated
  - Subsequently liver lesion found to be of no consequence (ie old scarring).
Case 4

- A fiercely independent patient has severe bilateral arm weakness secondary to ALS. She has stated on multiple occasions that she wishes to die when she is no longer able to live independently in her own home. She is now at that point.
  - How do you respond to her request?
  - What options do you have?
Case 4

- Is there a way to keep her in her residence?
  - Are there other public supports available?
  - Can she hire someone privately? Free room and board for assistance with care?

- Response may correlate with her respiratory status
  - If she is currently on BIPAP, stopping BIPAP could accelerate death (symptoms of dyspnea managed pharmacologically)
  - If there is no significant respiratory involvement, is MAID an option?
Medical Assistance in Dying

- Potentially an option
- But….
  - Would you all agree that she is in an advanced state of decline?
  - Would you all agree that her death is naturally foreseeable? (I would predict natural death likely much more than 1 year away, likely 2-3 years or more)
Definitions reminder

Grievous and irremediable medical condition is defined as:

a) They have a serious and incurable illness, disease or disability;

b) They are in an advanced state of irreversible decline in capability;

c) That illness, disease or disability, or that state of decline causes them enduring physical or psychological suffering that is intolerable to them and that cannot be relieved under conditions that they consider acceptable; and

d) Their natural death has become reasonably foreseeable, taking into account all of their medical circumstances, without a prognosis necessarily having been made as to the specific length of time that the individual has to live.
ALS and MAID

- I’m very doubtful that anyone would second guess “foreseeable natural death” when Dx is ALS
  - Average survival after onset of symptoms in ALS is 2-3 years.
  - An external accessor (ie coroner’s office) would not likely question foreseeable death in any patient with ALS
MAID discussions in ALS

- A large percentage of MAID conversations in clinic are just for information.
- Several patients die naturally from ALS, even if they have requested MAID.
  - Nov 1st reporting is looming!
Resources


- [www.als.ca](http://www.als.ca)
- [www.alsa.org](http://www.alsa.org)
Questions?