ALS and End-of-Life

Information for the Healthcare Provider

Review—what is ALS

• Amyotrophic Lateral Sclerosis
• Progressive neurodegenerative disorder
• Death of motor neurons cause progressive muscle weakness, atrophy and paralysis of the voluntary muscles
• End of life for ALS is respiratory failure due to diaphragm paralysis

Early End of Life Conversation

• ALS Center of Excellence at Nebraska Medicine
• Support Groups
• Care Service Consultations
Hospice Criteria for ALS

• Patients must fulfill one of the three following criteria to be considered to be in the terminal stage of ALS
  1) Patient should demonstrate critically impaired breathing capacity (Demonstrated by the following characteristics in the 12 months preceding initial hospice certification)
     1) Vital Capacity less that 30% of predicted
     2) DSypnea at rest
     3) Patient declines invasive ventilation; noninvasive ventilation used for comfort measures only

Hospice Criteria con’t

• 2) Patient should demonstrate rapid progression in ALS and critical nutritional impairment.
  – Rapid progression characterized as:
    a. Progression form independent ambulation to wheelchair to bed bound status
    b. Progression from normal to barely intelligible or unintelligible speech
    c. Progression from normal to pureed diet
    d. Progression from independence in most ADLs to needing major assistance by caretaker in all ADLs.

Hospice Criteria 2 con’t

– Critical nutritional impairment characterized by the following:
  • Oral intake of nutrients and fluids insufficient to sustain life
  • Continuing weight loss
  • Dehydration or hypovolemia
  • Absence of artificial feeding methods, sufficient to sustain life, but not for relieving hunger
Hospice criteria 3

• 3) Patient should demonstrate both rapid progression of ALS and life-threatening complications
  – Rapid progression as described in Criteria 2
  – Life threatening complications as characterized by the following:
    • Recurrent aspiration pneumonia (with or without tube feedings)
    • UTI
    • Sepsis
    • Recurrent fever after antibiotic therapy
    • Stage 3 or 4 decubitus ulcers

Hospice criteria considerations

• Co-Morbidities
  – Several listed but in particular to note—Dementia

   Important to remember that the two critical end-stage factors in determining the end-state of ALS
  1) Ability to breath
  2) to lesser extent the ability to swallow

Could this be the same disease?

• Limb onset
• Bulbar onset
• Respiratory onset
• Dementia onset/symptoms
End of Life Symptoms

- Psychosocial
- Mobility (skin)
- Constipation
- Presence of Feeding Tube
- Fatigue
- Cognition
- Pain
- Sleep Disturbance
- Sialorrhea
- Dyspnea

Psychosocial

- Common feeling expressed at end of life
  - Exhaustion
  - Fatigue
  - Fear
  - Anxiety
  - Increase burden on family
- Ensuring loved ones will be cared for in all aspects (Financial concerns)
- Meeting a goal or participating in a life event

Mobility Issues

- Assistive equipment plays a major role in the lives of persons with ALS to maintain independence
- Importance of ROM and positioning
- For the love of my power wheelchair—adaptations and ALS loan closet
- Fatigue and discomfort may contribute to more time in bed at end of life
Constipation

- Reduced mobility
- Fluid intake
- Meds (muscle relaxants, sedatives and anti-cholinergics)

Management
- Fluid intake
- Fiber
- Senna
- Bisacodyl suppository
- Magnesium citrate/Lactulose

Presence of Feeding Tube

- Timing
  - Dysphagia
  - Weight loss
  - Breathing
  - Time to eat meal
- Percutaneous or radiographic
- May improve QOL
- Unclear impact on survival
- Does stabilize weight/BMI

- Issues if not selected
  - Dehydration
  - UTIs
  - Aspiration Pneumonia
  - Quicker progression due to rapid weight loss
  - Usually more anxiety producing for family caregiver in regards to choking risks

Pain

- Muscle cramps
- Spasticity
- Musculoskeletal pain
- Joint contractures/frozen shoulder
- Skin pressure pain due to immobility
- Non-ALS pain
- Decubitus ulcers
Fatigue

- Fatigue may be a symptom of
  - depression
  - poor sleep
  - abnormal muscle activation
  - immobility
  - respiratory dysfunction

Fatigue management

- Screen for nocturnal hypoventilation
- Energy conservation
- Modafinil
- Amantadine
- Pyridostigmine
- Methylphenidate

Cognition Impairments

- There is now clear clinicopathological overlap between ALS and frontotemporal dementia, and with rigorous neuropsychological testing it may be possible to demonstrate mild cognitive impairment in up to 50% of patients.
- Although frank dementia is rare (and often an early feature when present), mild frontal lobe dysfunction might in theory impair executive function and influence decision making.
- Thus there may be a need to consider some decisions earlier on in the disease process, to allow patients to be fully involved.
- Estimates of cognitive impairment range from 10% to 75%.
- Fifteen percent of patients presenting with FTD later develop motor neuron degeneration.
Cognitive and behavioral impairment

ALS-FTD

- FTD, as defined by Neary et al. has insidious onset, gradual progression, altered social conduct, emotional blunting, and loss of insight. These criteria are required for the diagnosis of FTD, which is supported by neuropsychological abnormalities, language dysfunction, and poor self-care.
- Frontotemporal dysfunction with deficits in attention, cognitive flexibility, and word generation, with relative sparing of visuospatial function and memory.
- Changes in social interactions unrelated to a psychiatric condition.

Cognitive Impacts

- Extremely challenging for family caregivers, especially if mobility is still present
- Fewer interventions chosen/tolerated (i.e., feeding tube, non-invasive ventilation)
- Higher percent of ALS-FTD clients may need facility placement.

Sleep Disturbances

- Respiratory insufficiency with frequent awakenings due to nocturnal hypoventilation
- Anxiety and depression
- Inability to change position during sleep due to weakness
- Fasciculations and muscle cramps
- Dysphagia and aspiration of saliva
Sialorrhea

**Pharmacological**
- Glycopyrrolate
- Amitryptyline
- Transderm scopolamine patch
- Atropine 1% OP drops
- Myobloc

**Non-Pharmacological**
- MIE or Cough Assist
- Suction machines

Dyspnea and ALS—Noninvasive Ventilation

- Use nocturnally initially
- Traditional Bilevel machines
  - Set inspiratory and expiratory pressure
  - Back up rate
- Ventilators used non-invasively
  - Can do pressure or volume assisted breaths
  - “Smart” — target tidal volumes, adjusts pressure to reach it
  - Can use in sip/puff mode
  - Battery life
- CPAP not used
  - Weakness of oropharyngeal muscles
  - Increased burden breathing out against high pressure

Management of respiratory symptoms in ALS Orla Hardiman

- A significant minority of patients are unable to tolerate NIV.
- The mask may induce a sense of claustrophobia, and patients may have difficult acclimatizing to the sensation of increased inspiratory air pressure.
- Consider lorazepam at the initiation of NIV to help with relaxation.
- A variety of different types of masks are available.
- Factors that adversely affect the ability of patients to tolerate non-invasive positive pressure ventilators (NIPPV) include:
  - presence of bulbar symptoms with increased secretions
  - ability to manually adjust the mask
  - presence of cognitive impairment
Importance of Respiratory Therapist

- Assisting with Noninvasive Ventilation issues including mask fits and settings as disease progresses
- Assist hospice in educating person with ALS and their family on what the machine can and can not due for survival
- Education on MIE and secretion clearance
- Assisting with “terminal wean” from 24/7 noninvasive ventilation


- The fear of “choking to death” is on the mind of most patients suffering from ALS.
- We therefore performed a structured telephone interview with the relatives of 123 patients who were followed by the Wisdom Hospice, Rochester, NY. These data are compared with those obtained by a retrospective analysis of medical records of 56 ALS patients who were followed by the Wisdom Hospice, Rochester, NY.
- The data show that most ALS patients (Germany 88%, UK 88%) died peacefully, and no patient “choke to death”.
- The symptoms most frequently reported for the last 24 hours were dyspnea, coughing, anxiety and restlessness. Around half (55%, UK 52%) of the patients died at home.
- The main palliative measures in place during the terminal phase were: non-mechanical ventilation (21%, UK 0%), percutaneous endoscopic gastrostomy (G 27%, UK 14%), morphine (G 27%, UK 62%) and benzodiazepines (G 32%, UK 64%).
- The use of these palliative measures was judged to be beneficial by almost all relatives. These data support the hypothesis of a peaceful death process in ALS and should be communicated to patients and their relatives, at the latest after the onset of dyspnea, to relieve unwarranted fears.

Cause of death in 302 French patients with ALS

- Most patients (63%) died in a medical facility
- The most frequently reported cause of death was:
  - respiratory failure (77%)
  - including terminal respiratory insufficiency (58%)
  - pneumonia (14%)
  - asphyxia due to a foreign body (3%)
  - pulmonary embolism (2%)
- Ten per cent of patients died from other causes: post-surgical or traumatic conditions (5%), cardiac causes (5-4%), suicide (1.3%) and sudden death of unknown origin (0.7%)
- The cause of death could not be determined in 13% of cases (6% inside a medical facility and 7% outside)
Causes of death in a series of Italian ALS patients

- Between 2000 and 2008, 182 ALS patients (onset: spinal, 127; bulbar, 55; M/F: 1.6) were followed in a single ALS Tertiary Centre in Palermo, Sicily, Italy until death.
- Respiratory failure (terminal respiratory insufficiency, pneumonia) was the most frequent cause of death (81.3%), which included six cases (3.3%) who requested a terminal sedation.
- Sudden death and death during sleep accounted for by 6.0% and 6.6% of all deaths, respectively.
- Heart-related causes of death were relatively infrequent in our cohort, accounting for 7.1% of all deaths (i.e. sudden death: 6.0% and myocardial infarct: 1.1%).
- Patients (85.2%) died at home.
- The leading cause of death in ALS remains respiratory failure, followed by the sudden death and death during sleep.

End Stage Caregiver Issues

Countertransference issues exist with difficulties caregivers have in caring for ALS patients.

- Fear and isolation
- Disgust
- Anger and frustration
- Guilty
- Unappreciated or resentful
- Confronting own mortality
- Burnout!
- I LOVE THIS PERSON BUT I HATE THIS DISEASE!

High Caregiver Burden

- Krivickas (1997) study showed the primary caregiver of an ALS patient is spending a median 11hr per day caring for patient despite having home care assistance
- 42% reported feeling physically unwell
- 48% reported feeling psychological unwell
- Home care received by patients with ALS is often inadequate or too late to relieve the burden on placed on the family caregiver
Caregiver Quality of Life (QoL)

- Coco et al. (2005) confirmed that ALS has a negative impact on QoL of patients and caregivers, however caregivers QoL was not impacted by increased caregiving.
- Areas with highest impact was health (physical and psychological) as well as family for both patient and caregiver.
- Patients and caregivers who endorsed spirituality as a significant domain did have higher QoL.

Caregiver QoL

- Murphey et al. (2009) examined factors that could impact caregiver QoL.
- Again level of care was not a predictor of QoL.
- Social problem solving and spirituality were the best predictors of QoL.

Caregiver perspective on Final months

- Ganzini et al. (2002) found the most common symptoms in the last months of life for ALS patients included: difficulty communicating (62%), dyspnea (56%), insomnia (48%) and discomfort other than pain (48%).
- One third of caregivers were dissatisfied with some aspect of symptom management.
- Two thirds were enrolled in hospice—and those caregivers felt they died peacefully.
Supporting the Caregiver

- Respite and accepting care into the home earlier rather than later
- Managing expectations
- Education
- Low Tech/No Tech communication methods implemented

Supporting the Caregiver

- Developing interventions to teach ALS caregivers effective methods of problem solving (laying out possible scenarios and plans)---No one like surprises.
- Use of spirituality as coping if present.
- Caregiver support groups
- ALS Care Connections

How The ALS Association Can Help

- Provide in-services to hospices serving ALS clients
- Assistance with equipment through loan closet
- Be involved with family meetings when helpful
- Emergency respite funding if needed
- Connection to ALS Clinic team members at Nebraska Medicine

- The ALS Association Mid-America Chapter Programs:
The ALS Association

The ALS Association
Mid-America Chapter

(866) 878-2062
(402) 991-8788
stodd@alsa-midamerica.org
www.alsa-midamerica.org