The Final Days of ALS: perspectives for nursing

Christine Newell RN, Program Team Manager
Tamara Wells RN MN CNS
WRHA Palliative Care Program
Understanding

“ I’m lying in bed, Glistening beads of sweat have formed on my brow. My chest is heaving. Above me, quivering with anticipation is the focus of all my concentration. With one swift penetrating plunge, a flood of hot fluid rushes to the surface and finally, in a heap of spent flesh, I surrender. One more satisfied mosquito has had its way with me.”

Dennis Kay (1993)
Goals and Objectives

- To review the illness trajectory of ALS and how this relates to end of life.
- To develop an understanding of the patient/caregiver experience.
- To understand possible symptoms experienced related to end of life care for those with ALS.
- To develop an awareness of the pharmacologic and non-pharmacologic strategies used to manage these symptoms.
- To have the opportunity to talk about end of life care with ALS.
ALS

Two types of presentation

- Upper motor neuron involvement (Bulbar)
  - Difficulty with speech and swallowing
  - Incoordination
  - Slowing of movement
  - Stiffness/spasticity
  - Brisk reflexes
  - Excessive laughing or crying

- Lower motor neuron involvement (Spinal)
  - Weakness
  - Cramps
  - Muscle twitching
  - Muscle atrophy
Common Signs and Symptoms

- Shortness of breath
- Difficulty clearing secretions
- Dysphagia
- Sialorrhea
- Emotionally lability
- Weakness
- Muscle cramps
- Muscle twitching (fasiculations)
- Fatigue
- Pain to the limbs
- Sleep disturbance
What is Not Lost

- Sensation
- Urinary sphincter control
- Bowel Sphincter control
- Ocular Muscle
  - Communication
Challenges of Diagnosis

- Diagnosis of exclusion
- No definitive markers or tests
- Testing non-specific
Psychosocial Aspects: Patient Experience

- **Isolation**
  - From family, friends
  - The world, nature
  - Quality of Life meanings

- **Loss of self**
  - Physical losses
  - Role Losses

- **Treatment decisions**

- **Living with the unknown**
Patient Experience

- Coming to terms
- Quality of life (McDonald 2014)
  - Represented as “the spirit” of the patient
  - QoL is realistic despite profound losses
- Preserving hope (with caregiver support)
  - Realistic expectations
  - Drawing on inner strength
  - Support systems (family, health care professional, psychosocial supports)
- Shifting focus from ALS to sharing life together
- Communication
- Inclusion of family
Caregiver Experience  (McDonald 2014)

- Isolation
  - Family, friends, occupation

- Health changes
  - depression

- Loss of sense of self
  - Resentment, guilt

- Stress
  - Caregiver stress
  - Role stress
    - Wife to nurse
    - Relationship stress
    - Lack of occupation
    - Role reversals
Nursing Experience (ALS Society of Canada)

- Emotional challenges
  - Personal conflicts
  - Fear

- Ethical challenges
  - What treatments are right for which people?
  - What is Quality of Life?
  - Who is to decide?

- Frustration
  - Inability to fix the problem(s)

- Isolation
  - Am I the only person that feels this way?
The Nursing Role

- Bearing witness
  - Open ears, open heart
- Being open to conversation
- Facilitating the right people to have the right conversation
  - You are not expected to have all the answers
- Symptom assessment
- Symptom management
- Self care
Challenges to patient care

- Emotional lability
- Communication struggles
- Balancing time and patient need
- Promoting developmental independence adaptations
- Anticipation of future problems
- Helping family members with role changes
- Grief
Communication

- Numerous challenges
- Requires time
- Use different technologies
  - Confusion
  - Safety issues (i.e., call light systems, back up plans)
  - More than one mode
  - Loss of non-verbal communication
  - Proactive discussion about future communication abilities
What to Expect

- Progressive functional decline
  - May be slow or rapid
  - Very individual progression
  - Until the end when death is often sudden
- Worsening of symptoms
- Development of new symptoms
- Time to re-evaluate and have discussions
  - At each stage communication, communication, communication
  - What are the goals?
The Momentum of Change

- Often death is very sudden
  - Death often occurs at night
  - 40% died within 12 hours of change in condition being noted.
- Teams caught unprepared and unaware
- Families caught off guard
- Not the same scenario for those with BiPap or invasive ventilation
  - Bipap can delay the onset of the final days but not its character
  - Invasive ventilation leads to a locked-in state

(Sykes, 2006)
Case Presentation

- Mr. B
  - Mid 50s
  - Was working as a professional
  - Bulbar presentation
  - Fairly healthy other than ALS
  - Lived alone
  - Had supports
  - Hired own help
  - Tech savvy
  - Communicated verbally and with technology
  - Liked to be in control of his life
Respiratory Failure: Hypoventilation

- Most common cause of death in ALS
  - Neuromuscular failure
- Caused by hypoventilation
  - Changes to the inspiratory and expiratory muscles
- Can be multifactorial
  - Aspiration
  - Pneumonia

**Definition:**
- Inadequate alveolar ventilation in relation to metabolic demands. Caused by altered pulmonary mechanic or in the neurologic control of breathing. CO2 removal does not keep up with production = hypercapnea. (Brashers, 2006)
Hypoventilation

- **hypercapnea**
  - May go undetected
  - Can only detect with ABG
    - pCO2 >45mmhg
  - Adaptation
    - Symptoms progress and CO2 rises
    - Oxygenation may decrease
    - Sedation, somnolence, confusion

(Lyall & Gelinas, 2006)
Ventilation

- **Invasive**
  - Personal choice
  - Discussion about end points
  - Not a cure but a management strategy
  - Own set of problems
    - Equipment, infection, training and support

- **Non-Invasive**
  - Cpap/BiPap
    - Personal choice
    - Safety concerns may be higher related to loss of independent function, lack of alarms
    - Not a cure but a management strategy
Communication

- Don’t assume that the absence of question reflects an absence of concerns
- Upon becoming aware of a turning point in illness; it would be very unusual not to wonder:
  - “How long do I have?”
  - “How will I die”
- Waiting may result in missed opportunities to address concerns
- Consider exploring preemptively
Goals of Care/Advanced Care Planning

- For every intervention or non-intervention there is a response
- Advance Care Planning (ACP; WRHA)
  - The overall process of dialogue, knowledge sharing, and informed decision-making that needs to occur at any time when future or potential life threatening illness treatment options and Goals of Care are being considered or revisited.
- Goals of Care
  - The intended purposes of health care interventions and support as recognized by both a Patient or Substitute Decision Maker and the Health Care Team
Substitute Decision Making

- It is not our role to determine patients’ goals of care
  - How to make these decisions must be identified early in the disease process
- Family proxy or other designated proxy role
- How do you help with this?
  - Know the patient
    - What gave them QoL, who are they now, who were they before?
  - Ask the family
    - If mom had left a note in her pocket from a year ago when she was well, what would it say to do now?
    - If your dad was standing here looking at this situation what would he tell you to do
The Case for Palliative Care

- Identified that palliative care intervention improves QoL (Bede, et al., 2011)
  - Unfortunately, not happening in a consistent manner
  - Barriers include:
    - When to initiate
    - Balancing hope and reality
    - Cognitive changes
    - Coordination of care

- Benefits: (Bede, et al., 2011; Blackhall, 2012)
  - Decreased symptom burden increases QoL and can extend life
  - Can help communication about treatment decisions
Case Presentation

- Mr. B
  - Chose no form of ventilation
  - Realized that his idea of independent function needed to change
  - Became a bit more withdrawn but developed a strong faith and trust in his new care team
  - Loss of muscle function began to speed up
  - Had PEG tube
  - Was no longer eating orally, meds via PEG
Symptom Management

- Dyspnea
- Choking
- Sialorrhea
- Anxiety
- Other
Medication routes: Sublingual or Buccal

- **Advantages:**
  - Easy to use
  - Rapid absorption

- **Limitations:**
  - Maximum volume of 1 ml
  - Not all medications are well absorbed
  - Swallowing may delay onset or reduce effectiveness
Subcutaneous Route

Advantages:
- Can be used intermittently
- Indwelling catheter comfortable
- Useful alternative to IV for some medication and fluid

Limitations:
- Local tissue reactions / potential for infection
- Moderate cost of supplies / requires policy and staff education
- Contraindicated if gross edema
Transdermal Route

- **Advantages:**
  - Easily applied
  - Useful if person is confused or delirious (can be applied to inconspicuous location)

- **Limitations:**
  - Slow release: does not provide rapid relief of acute symptoms
  - Patch adherence and changes to skin permeability affect drug absorption
Fentanyl Patch Safety

- Do NOT use if opioid naïve
- Not for pain crisis
  - Indicated for people living with chronic, moderate to severe pain that is well controlled using stable doses of opioid
  - Fentanyl 12 mcg patch = minimum 45 mg oral morphine per day
  - Takes 12 - 24 hours to reach therapeutic drug levels
  - Breakthrough medication is required
Intranasal Route

Advantages:
- Accurate dosing, easy to administer
- Rapid absorption

Limitations:
- Some medications irritating
- Only used with certain meds
Rectal Route

- **Advantages:**
  - Easy to use
  - Alternative if other routes not feasible

- **Limitations:**
  - Inappropriate if the person has diarrhea or bowel incontinence
  - Can create discomfort
  - Invasive / can impair sense of dignity
Dyspnea (Shortness of Breath)

- “an uncomfortable awareness of breathing”

Subjective
- Even if they don’t look short of breath they still can feel short of breath

Assessment
- Ask them
- Non-verbal cues
  - Fidgety
  - Brow furrowed
  - Eyes darting
  - More interactive, call bells etc.
  - Increased accessory muscle use
Dypsnea

- Non-pharmacologic
  - Fan blowing gently on the face
  - Pacing activity
  - Balancing feeding and activity
  - Calm reassurance
  - Distraction
  - Mouth care
  - Oxygen for some
  - Manage constipation/bowel habits
Dyspnea

- Pharmacologic
  - Opioids:
    - Morphine 2.5-5mg orally or 1.25-2.5mg subcut
    - Hydromorphone 0.5-1.0mg orally or 0.25mg-0.5mg subcut
      - Note: start low and go slow
      - Above dosing for opioid naïve pts
      - Long term opioid users would need dosing based on underlying usage.
  - Medications should be given regularly and prn
  - Lorazepam
Subcutaneous Morphine In Advanced Cancer

Bruera et al. JPSM 1990; 5:341-344
Anxiety

- Common response to daily stressors
- Common co-symptom with dyspnea
  - Must be taken seriously
  - Assess regularly

Assessment
- Ask
- Talk to the family
- Look for non-verbal cues
  - Furrowed brow
  - Call bells
  - Withdrawal
  - Eyes darting
- Increased WOB (accessory muscles, frog breathing)
Anxiety

- **Non-pharmacologic**
  - Reassurance
  - Distraction
  - Calm presence
  - Pace activity
  - Know your patient’s triggers

- **Pharmacologic**
  - Lorazepam
  - Opioid adjunct
    - Morphine, hydromorphone, fentanyl
Choking

- Often a symptom with bulbar presentation
  - Related to loss of head and neck muscle function
  - Hyperreflexia
  - Often an occurrence with oral secretions and feeding
- Sudden and Severe
- Can lead to other symptoms
  - Anxiety
  - Shortness of Breath

(Sykes, 2006)
Choking

- Care with feeding
  - Upright
  - Don’t rush
  - Appropriate textures and foods
  - Communication
- Manage oral secretions
  - Glycopyrylate
  - Scopolamine
  - Other anticholinerics
- Good oral hygiene

- Management of chest secretions
  - Guafenesin
  - Inhaled sterile water
- Positioning
  - What helps the patient to manage
Secretions

- Pooling related to dysphagia
  - May be copious oral
  - May be thickened chest

- Management
  - Determine the cause/what are the goals of care
    - For sialorrhea:
      - Glycopyrrolate
      - Scopolamine
    - For chest: quafenesin to thin
      - When might you want to consider this should be stopped?

(Wagner-Sonntag, & Prosiegel, 2006)
Pain

- Incidence 19-80%
- Due neurological changes
- Due to lack of control over positioning
- Not usually anticipated to develop closer to death
- Death itself is not painful
Pain Management

- Non-pharmacological
  - Positioning
  - Distraction
    - Music
    - Television
    - Technology
    - Family
  - Use of therapeutic surfaces
  - Management of other symptoms
  - Heat/cold
  - Massage
  - Attention to psychosocial concerns
Pharmacological

- **Opioids**
  - Morphine 2.5-5mg orally or 1.25-2.5mg subcut
  - Hydromorphone 0.5-1.0mg orally or 0.25mg-0.5mg subcut
    - Note: start low and go slow
    - Above dosing for opioid naïve pts

- **Neuropletics**
  - Gabapentin

- **Other**
  - Acetaminophen, NSAIDS
Case presentation

Mr. B.
- Decreasing tolerance for activity
  - In bed more, not tolerating turns
- More difficulty with secretions
  - Expectorating, feelings of fullness in chest, sialorrhea
- More episodes of feeling short of breath and anxious
  - Chicken egg scenario
  - More difficult to meet expectations
  - Needing more support
- Less accessory muscle use
  - Coping with respiratory events more challenged
<table>
<thead>
<tr>
<th>Symptom</th>
<th>Suggested First Steps</th>
<th>Next Steps</th>
<th>Third Step</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shortness of Breath</td>
<td>Block Care activity.</td>
<td>may start to feel anxious</td>
<td>continues to be short of breath and anxious Fentanyl 25-50mcg given intranasally.</td>
</tr>
<tr>
<td>Becomes short of breath with position changes.</td>
<td>Assess this feeling regularly</td>
<td>Remain <strong>calm</strong> and provide reassurance</td>
<td>May repeat in 10 minutes if symptoms and situation not resolving</td>
</tr>
<tr>
<td></td>
<td>Avoid positioning and transfer during feeding and for 15-30 minutes after feeding.</td>
<td>hydromorphone 0.5mg, given sublingually for shortness of breath. (normally works in less than 5 minutes) Nursing may provide by PEG if oral secretion copious</td>
<td>Have someone stay at bedside</td>
</tr>
<tr>
<td></td>
<td>If he indicates that he is short of breath when being positioned stop and either:</td>
<td>Medication may take 5 minutes or longer to start working.</td>
<td>If situation non-resolving contact the PCP</td>
</tr>
<tr>
<td></td>
<td>return him to sitting position, turn him lower the lift, get him off the commode.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td><strong>Stay calm</strong> and provide a reassuring presence.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Fan blowing gently on face may help.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>often feels anxious when short of breath</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anxiety</td>
<td>Inform patient of care and block care.</td>
<td>may need a prn ativan.</td>
<td>may require intranasal fentanyl.</td>
</tr>
<tr>
<td>---------</td>
<td>---------------------------------------</td>
<td>-----------------------</td>
<td>---------------------------------</td>
</tr>
<tr>
<td></td>
<td>Ask patient routinely while awake if he feeling anxious.</td>
<td>Provide supports and comfort, such as music, tv, reassuring touch,</td>
<td>Have a HCA stay and provide calm reassurance</td>
</tr>
<tr>
<td></td>
<td>If he is feeling anxious reassure patient and stay with him until he settles</td>
<td>Continue to reassess if his anxiety is resolving</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Consider environmental supports and distraction. Such as music, tv, quiet, gentle touch, a familiar presence.</td>
<td>If anxiety continues patient may begin to feel short of breath.</td>
<td>Provide prn hydromorphone</td>
</tr>
</tbody>
</table>
Final Hours

- Encourage communication with resident as if they were awake and able to communicate,
  - even when the resident is unconscious they still be able to hear you

- Family may have things that they want to say
  - Helps with closure, encourages connectedness

- Encourage time with their loved one after death has occurred
Final Hours: what you probably won’t see

- Decreased or no urine output
- Reflexive actions such as the: grasp, sucking, or rooting reflex
- Inability to take anything orally
- Mostly sleeping
- Mottling of the extremities
- Breathing changes

*Often your best cue for what is happening is your ability to listen to the patient*
What you may see

- Careful attention to subtle patient changes can provide the clues to what will be happening next
  - Sudden changes in level of consciousness
  - Sudden onset of symptom concerns
  - Seminal event
  - Lack of awakening
Conclusion
Questions Comments
Resources

- Blachall, LJ. Amyotrophic lateral sclerosis and palliative Care: where we are and the road ahead. Muscle & Nerve 2012, 45, 311-318. DOI 10.1002/mus.22305
- Canadian Virtual Hospice www.virtualhospice.ca
- WRHA Palliative Care Program 204-237-2400