Palliative Care in Amyotrophic Lateral Sclerosis (ALS)

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Objectives

1. Recognize signs and symptoms of ALS at various stages

2. Know about interventions that can prolong life and/or increase quality of life

3. Understand the role of an interdisciplinary team

4. Discuss the resources available in your communities
Outline

- What is ALS?
- Symptoms
- Interdisciplinary Team
- Resources

Disclosure

No conflict of interest to declare
Case

- "Fred": 65 y.o. man, previously healthy, no family history, no medications
- 6 months ago: right foot drop
- 3 months ago: left hand fasciculations
- 1 month ago: right arm atrophy
- 2 weeks ago: progressive dysphagia to solids
Case Continued

- Decreased reflexes in upper limbs, increased reflexes in lower limbs, fasciculations seen, right leg atrophic, spastic gait, no sensory loss
- CT head: no obvious mass or bleed; lab investigations unremarkable
- Urgent referral to neurologist, clinical diagnosis of ALS made
What is ALS?

- “Lou Gehrig’s Disease” in Canada/US, “Motor Neurone Disease” in UK/elsewhere
- Etiology unknown
What is ALS?

- Progressive, terminal neurodegeneration, loss of upper and lower motor neurons
- Eventual paralysis of voluntary muscles and inability to swallow, speak and breathe
- Spares eyes, heart, bowel, bladder and sexual organs
- Can have cognitive and behavioural changes, mimicking fronto-temporal temporal dementia
The Numbers

- Age of onset 40-70 (average age 55)
- Affects men and women
- Incidence 2 per 100,000 per year
- 2500-3000 Canadians currently live with ALS
Kingston ALS clinic (Dr. J. Wee) follows 40 patients per year, with 12 new consults per year

Life expectancy typically 2-5 years
- 20% live over 5 years, 10% live over 10 years

Mortality 2 per 100,000 per year
Classification

- Classical (sporadic) ALS: 90% of cases
- Familial (genetic) ALS: 5-10%
  - 20% of these related to mutation in copper zinc superoxide dismutase (SOD1) on chromosome 21
- Region of onset: limb vs. bulbar (mouth/face/throat)
- Not contagious
Diagnosis

- No specific lab or investigation
- Diagnosis made by neurologist (early referral!)
- Revised El Escorial criteria:
  - Lower motor neuron (LMN) degeneration seen clinically, by EMG or biopsy; AND
  - Upper motor neuron (UMN) degeneration seen clinically; AND
  - Progressive spread of signs or symptoms within a region or to other regions

Elman 2011
**Signs**

- Usually onset is focal and asymmetric
- Can begin with limb wasting and weakness, or changes in speech and swallowing
LMN Signs

- Muscle weakness and atrophy (including diaphragm)
- Fasciculations
- Muscle cramps
- Hyporeflexia
- Flaccidity
UMN Signs

- Dysarthria (impairment of muscles of speaking)
- Dysphagia (difficulty swallowing)
- Dyspnea
- Pseudobulbar affect (inappropriate uncontrollable laughter or crying)
- Sialorrhea (excessive salivation)
- Hyperreflexia
Outline

- What is ALS?
- Symptoms:
  - Dysarthria
  - Dysphagia
  - Dyspnea
  - Sialorrhea
  - Pseudobulbar Affect
  - Pain
- Interdisciplinary Team
- Resources
Fred was referred to interdisciplinary team

Frustrated, anxious, fearful

Team took time to explain how they could help
1. Dysarthria

- Speech affected by weakness or paralysis of muscles of lips, tongue, jaw, soft palate and larynx
- Slurring, hoarseness, breathy voice
- Isolating and frustrating; loss of control if misunderstood or ignored
1. Dysarthria Management

- Early SLP and OT referral
- Augmentative and Alternative Communication (AAC):
  - Alphabet board
  - Lightwriter
  - TTY
  - ETRAN board
1. Board with Words

Permission of patient obtained by Dr. I. Stewart
1. Communication Tips

- Position face to face
- Establish reliable “yes” and “no”
- Do not interrupt or try to finish sentences unless asked by a patient to do so
2. Dysphagia

- Weakness of lips, tongue, masseter, soft palate or esophagus
- Coughing, difficult chewing and swallowing, reduced airway protection
- Drooling, malnutrition, dehydration or aspiration (increased risk of pneumonia)
2. Dysphagia Management

- **SLP:** Swallowing assessment and advice
- **Dietician:** Texture modification, supplements and eating strategies
- **OT:** Adapted feeding tools, arm supports, etc.

- **Hypermetabolic state:**
  - Loss of muscle mass, decreased intake, increased energy cost of activities
- **Assess bowel function and monitor weight**
2. Tube Feeding

- When to consider?
  - Eating becomes exhausting
  - Nutritional goals not being met
  - Oral intake time-consuming

- G-tube, J-tube, PRG tube

- Tube feeding does not eliminate risk of aspiration

- Can still have oral feeds if tolerated
2. Tube Feeding Continued

- Timing of PEG depends on breathing function, coordinated by neurologist and respirologist
- Early discussion of wishes and goals of care
- Tube feeding improves both quality and quantity of life
3. Dyspnea

- Shortness of breath, fatigue, anxiety, claustrophobia and insomnia
- Speaking or eating can be difficult
- Hypoventilation worse during sleep
- Increasing CO₂ levels → headaches, somnolence, nausea
- Respiratory failure is most likely cause of death
3. Dyspnea

- Monitor pulmonary function (e.g. vital capacity, cough ability)
- Measuring ABG helpful to guide prognosis and see if O₂ warranted
- In CO₂ retention, target SpO₂ to 88-92%
- Clarify treatment goals with O₂
- In hypoventilation, BiPAP improves quality of sleep (including REM sleep)
3. Dyspnea Management

- Options:
  - Medical management
  - Non-invasive ventilation (BiPAP)
  - Tracheostomy and long-term invasive mechanical ventilation
3. Respiratory Secretions

- Secretions compromise airway, add to discomfort and panic
- Insufflator/exsufflator (Cough Assist) device
- Suctioning
3. Dyspnea Management

- BiPAP improves quality (and maybe quantity) of life
- Invasive mechanical ventilation appropriate if goal is long-term survival, with supports
- Advance directive discussion is key
- Emphasize that “choking to death” is almost unheard of in ALS
4. Sialorrhea

- Normal saliva production but difficulty clearing due to impaired muscle function
- Anterior pooling of secretions and poor lip seal
4. Sialorrhea Management

- Anticholinergics:
  - Atropine, amitriptyline, scopolamine patches (S/E xerostomia and constipation)

- Good oral hygiene

- Portable suction device

- Botox injections into parotid or irradiation of salivary glands (Level B evidence) if medically refractory

- Tracheostomy if choking is life-threatening

ALS Society of Canada, Miller 2009
5. Pseudobulbar Affect

- Emotional lability:
  - Uncontrolled / inappropriate laughter or crying

- Due to lost inhibition of limbic motor neurons

- Seen in 50% of ALS patients

- Can be associated with frustration, social anxiety and social withdrawal
5. Pseudobulbar Affect Management

- Screen for depression
- Discuss social management
- Consider TCAs, SSRIs, valproate or lithium
- Neurodex:
  - Combo of dextromethorphan and quinidine
  - Level B evidence of benefit
  - Under investigation for safety in the U.S.
6. Pain

- Muscle weakness, stiffness, immobility
- Inability to maintain spinal posture
- Neuropathic pain from entrapment or positioning
- Muscle cramps
- Jaw spasms and laryngospasm
- Constipation
6. Pain Management

- Positioning and PT referral:
  - Stretching, repositioning, passive movements to prevent stiffness

- Medications:
  - NSAIDs, opioids, anticonvulsants, TCAs
  - Consider baclofen
  - Quinine for spasms no longer recommended
  - Insufficient data re specific treatment of cramps or spasms in ALS (Level U – unknown)

ALS Society of Canada, Miller 2009
Fred thanks you for the info

“Is there any treatment?”
Riluzole

- Riluzole prolongs trach-free survival but not shown to help quality of life

- American Academy of Neurology:
  - 4 RCTs show 2-3 month benefit in survival
  - 5 cohort studies showing up to 21 month benefit

- Cost $900/month, not in ODB database

- Prescribed 50 mg PO BID

ALS Society of Canada, Bedlack 2010, Miller 2009
Riluzole Continued

- Mechanism unclear
  - ? inhibits glutamate release

- Generally well tolerated, some nausea and fatigue

- Other drugs have no evidence in changing disease course:
  - Lithium
  - Vitamin E
  - Acetylcysteine
  - L-methionine
  - Selenium
  - Creatine
Outline

- What is ALS?
- Symptoms
- Interdisciplinary Team
- Resources
Interdisciplinary Team

- Goals are to provide information, promote function and independence, provide hope, conduct ongoing assessments, act as advocate.
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Resources

- ALS Society of Canada: [www.als.ca](http://www.als.ca)
  - Resources for patients/families and healthcare professionals
  - Local Kingston chapter no longer in operation

- Canadian ALS Research Network: [www.alsnetwork.ca](http://www.alsnetwork.ca)

- ALS Association (U.S.): [www.alsa.org](http://www.alsa.org)

- Canadian Hospice and Palliative Care Assoc.: [www.chpca.net](http://www.chpca.net)
ALS Clinic (Kingston)
The Adult Neuromuscular Clinic (Dr. J. Wee)
St. Mary's of the Lake Hospital Site
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Tel: 613-544-1894
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- Operates once a month
- Follows 40 patients/year, incl. 12 new consults
- Patients referred upon diagnosis
- Patients unable to cope at home may be CCC candidates

Wee 2012
How to Improve?

- ALS Society branch no longer in operation in Kingston
- Need for Augmentive Adaptive Communication (AAC) services
  - Funding removed by MOH from Kingston ~10 years ago
- Need for hospice care for end-of-life!
- Respite beds, ventilator beds
Fred started on Riluzole and continues to follow-up with the ALS team

Declines slowly over the next 2 years

Opts for G-tube and BiPAP
Eventually no longer able to tolerate BiPAP

Had decided previously not to pursue invasive mechanical ventilation

Opts to stay at home with family supports, CCAC and visiting physician

Dies at home 3 years after diagnosis, comfortably, from respiratory failure
Summary

- ALS is a progressive, terminal disease
- Early referral to a neurologist for diagnosis, and to an interdisciplinary ALS team is key
- Many symptoms have effective palliative treatments
- Discuss goals of care early, especially around feeding and breathing
- Always help patients maintain hope
Objectives Revisited

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References


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