

## Questions to be Included in the Canadians ALS Guidelines

### Communication

- AAN How should a physician tell patients that they have ALS?
- New What is the best timing for discussions around life sustaining therapies and goals of care, (feeding tubes, Invasive ventilation, etc)?

### Disease Specific Treatment

- AAN What is the effect of riluzole on slowing the disease process or prolonging survival in ALS?

### Respiratory Management

- What is the best respiratory management of ALS? (Two main categories: respiratory insufficiency and airway clearance)
- AAN What pulmonary tests should used to detect and monitor respiratory insufficiency?
  - New How frequently should respiratory function be assessed?
  - New When should NIV be initiated? (plus statement about wait times)
  - EFNS How are bronchial secretions best treated in ALS?
  - New What are the indications for using oxygen in patients with ALS? Oxygen statement (is best practice to avoid oxygen in patients with ALS)
  - New What are the indications for LVR and cough assist in ALS? How often should it be used?
  - New What are the indications for tracheostomy in ALS (also consider referring people to chronic vent guidelines for transitioning to IMV from NIV)?
  - New Is diaphragmic pacing effective in ALS?

### Nutrition management

- AAN When is enteral nutrition indicated in ALS? (How is nutrition monitored, percentage of weight loss, aspiration, respiratory status) How should aspiration risk be assessed?
- New What is the maximum allowable delay for REG/RIG insertion, after it has been clinically recommended? (consensus statement attached to one)
- New What is the recommended procedure for feeding tube insertion? (endoscopic/fluoroscopic/inpatient etc)
- New What type of supplements/enteral feeds should be suggested (high fat/high protein statement)? (TPN comment?)
- New Should hyperalimentation be used in ALS?

### Dysarthria Treatment

- AAN What treatments for dysarthria optimize communication in ALS?

### Symptom Management

- AAN What are the most effective treatments for sialorrhea?
- AAN What pharmacologic measures reduce pseudobulbar affect?
- AAN What interventions reduce spasticity? (exercise included)
- AAN What interventions reduce cramps?
- AAN What pharmacologic interventions reduce depression (interactions with riluzole)?
- AAN What pharmacologic interventions reduce anxiety?
- AAN What pharmacologic interventions reduce insomnia?
- AAN What pharmacologic interventions reduce fatigue?
- New What is the prevalence and type of pain in ALS patients?

New What are the most effective treatments for fasciculations?

### **Palliative Care**

What is best end of life care in patients with ALS (as overarching question). Would include statement about requests for hastening death/physician assisted death.

AAN What treatments reduce dyspnea in the terminal phase of ALS?

AAN Do hospice care, spiritual interventions, or advance directive improve quality of life in the terminal phase of ALS?

AAN What is the optimal method of withdrawing both non-invasive and invasive ventilation in ALS?

New When should palliative care be offered?

New What bereavement counseling should be offered to caregivers of deceased patients with ALS?

### **Multidisciplinary Care**

AAN Does multidisciplinary management improve outcomes?

New What are the necessary components of multidisciplinary care?

New Algorithm/flow chart to potentially be designed [What measures (spirometry (including what exactly), oximetry, bloodwork, ALS-FRS-R, weight, etc) should be routinely monitored, and how often]

New How frequently should patients be seen in the multidisciplinary ALS clinic?

### **Cognitive Impairment**

AAN How is cognitive or behavioral impairment in ALS diagnosed?

AAN What treatments are effective for cognitive or behavioral impairment in ALS?

### **Safety**

#### *DVT surveillance*

EFNS What is the risk of DVT in patients with ALS?

EFNS Should DVT prophylaxis be considered in patients with ALS?

New What is the recommended safety monitoring for patients on riluzole?

### **Health Promotion**

#### *Exercise (limit to clinical studies)*

New What level and type of physical exercise is recommended in patients with ALS?

#### *Medication Alignment/Discontinuation*

New Should medication reviews occur regularly to ensure unnecessary medications have been discontinued? (can't limit to clinical)

New Should statins be discontinued in patients with ALS?. (limit to clinical)

### **Caregiver Support and Caregiver Health**

EFNS What is the burden of care for ALS caregivers? (Impact/caregiver burnout/respite/monitoring)