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Hospice Palliative Care Program
Symptom Guidelines

Amyotrophic Lateral Sclerosis (ALS)

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Rationale

This guideline is adapted for inter-professional primary care providers working in various settings in Fraser Health, British Columbia and the Fraser Valley Cancer Center and any other clinical practice setting in which a user may see the guidelines as applicable.

Although ALS creates many unique challenges, basic principles of palliative care support comfort and dignity.

Scope

This guideline provides recommendations for the assessment and symptom management of adult patients (age 19 years and older) living with advanced Amyotrophic Lateral Sclerosis (ALS). This guideline does not address disease specific approaches in the management of ALS.

While most cases of ALS occur sporadically, approximately 10% of cases are familial and 2% result from a specific gene defect on chromosome 21.[1-4] ALS has a world wide incidence of approximately 6 to 8 per 100,000; has greater incidence in males than females and usually occurs in mid life.[1, 2, 5, 6] Average survival from diagnosis is about 3 to 5 years,[2, 4, 6, 7] with a range of 6 months to 20+ years[4]. Survival time is less with bulbar onset (median 1 year) than limb onset (median 2.5 years).[1] Survival time is generally prolonged in younger patients.[2] Fasciculations (muscle twitching) are a prominent feature of ALS[1] in co-existence with muscle atrophy, weakness and cramps.[2, 4, 6]

Definition of Terms

ALS (also known as Lou Gehrig's disease) is a progressive fatal type of motor neuron disease resulting in spasticity, diffuse muscular atrophy and weakness.[4, 7] ALS is the most common type of motor neuron disease (MND).[1, 6] ALS was named according to its clinical picture (muscle wasting is amyotrophy) and pathological finding (hardening of the lateral aspects of the spinal cord secondary to gliosis of the pyramidal tracts is lateral sclerosis).[2]

Standard of Care

1. Background
2. Assessment of ALS symptoms
3. End-of-Life Discussions
4. Symptom Treatment

Recommendation 1 Background

New Treatment Approaches:

Research has linked ALS with high levels of glutamate, the primary excitatory neurotransmitter in the central nervous system. There is a theory that glutamate accumulates in nerve cell synapses, eventually destroying the nerve cell. Riluzole, the first drug specifically to treat ALS, inhibits the release of glutamate.[2, 3, 5, 8] It has been shown to extend the survival time of individuals with ALS.[4, 6]

Riluzole has been approved for use in the United States and has conditional approval in Canada. It can be obtained from the Vancouver Coastal ALS Centre or from Neurologists at the EMG Lab at Vancouver General Hospital. Recommended dose for riluzole is 100mg daily[5] divided into two 50 mg doses.[1]

Background

In North American society, which prizes independence and vitality, a diagnosis of ALS can be devastating.[4] Many people are uncomfortable with the profound physical changes of a disease that generally leaves a person mentally intact. The patient is often very distraught over diminishing quality of life, which raises ethical issues regarding aggressiveness of treatment, ventilation, Do Not Resuscitate (DNR) orders and assisted suicide.

Few diseases stimulate such a response of total pain. The patient experiences multiple losses such as mobility, speech, the ability to eat and drink, independence, relationships, personal fulfillment provided by a job or recreational activities. The frustration and loss of control may lead to anxiety, anger, depression and controlling behaviour. Emotional lability and bouts of crying are common. Up to 50% of ALS patients have pseudobulbar effect, pathological uncontrolled bouts of laughing or crying not concordant with their mood.[1, 4, 8, 9]

Families, friends and caregivers may be disturbed by the physical changes and/or perceive the patient's behaviour to be unreasonable or demanding.[4] People often visit less frequently thus increasing the loneliness and isolation the patient feels.

Communication and safety can become key concerns. Impulsive behaviour may contribute to safety issues.[8] There are many devices available to facilitate these important aspects of life. Referrals to occupational therapists in the local health area or through the ALS team are very beneficial.[5]

Throughout the illness, patients and families will require support from their neighborhood and community, and many health care providers and agencies.[1, 7, 9] Key resources are the Vancouver Coastal ALS Centre, a provincial resource which provides diagnosis, ongoing medical and multidisciplinary team access. The ALS Center is located at GF Strong Rehabilitation Centre. Another key resource is the ALS Society, which provides information, support, and equipment loan (i.e., electric wheelchairs, suction machines) to ALS patients. Their website has links to other ALS organizations and information for patients, families and health care providers.

Recommendation 1 Background continued...

Vancouver Coastal ALS Centre 1-604-737-6320

ALS Society of BC 1-604-685-0737 or 1-800-708-3228 or www.alsbc.ca

ALS Society of Canada www.als.ca

"The psychic burden of caregivers sometimes exceeds that of the patient."[2, 6, 9]

Recommendation 2 Assessment of ALS Symptoms

Ongoing comprehensive assessment is the foundation of effective symptom management for patients with Amyotrophic Lateral Sclerosis, including interview, physical assessment, medication review, medical and surgical review, psychosocial review, review of physical environment and appropriate diagnostics.

Identifying the underlying symptoms for patients with a diagnosis of ALS is essential in determining the interventions required (*see Table 1*).

- ALS often starts focally with muscle weakness or wasting, eventually spreading bilaterally affecting peripheral and cranial nerves and usually sparing bowel, bladder, sensory and cognitive functions.
- Brain stem involvement (bulbar ALS) produces dysarthria (slurred speech), dysphagia (swallowing problems), sialorrhea (drooling)[4] and aspiration.[1] It occurs in 20 to 30% of ALS cases[8], especially in older women.[2, 6, 9]
- Eye movement is usually preserved which may be useful in communication.[2, 4-6]
- Respiratory insufficiency is one of the most critical issues for the majority of patients with ALS.[7]

Table 1: Symptoms Due to ALS:[2, 6]

Directly	Indirectly
Weakness and atrophy	Psychic disturbances
Fasciculations and muscle cramps	Sleep disturbances
Spasticity	Constipation
Dysarthria	Drooling
Dysphagia	Thick mucous secretions
Dyspnea	Symptoms of chronic hypoventilation
Pathological laughter / tearfulness	Pain

Recommendation 3 End-of-Life Discussions

End-of-life discussions are an integral part of care for patients living with ALS and their families. This document does not provide a comprehensive guide for completing Advanced Care Planning for ALS.

Advance Care Planning is a process by which people can think about their values regarding future health care choices, hear medical information that is relevant to their health concerns, communicate wishes and values to their loved ones or agents, and document their choices so the decisions are available to health care providers wherever that person receives care.

For more information on *Advance Care Planning: Let's Talk,* view the information at:
www.fraserhealth.ca/HealthInfo/AdvanceCarePlanning
Or contact **advancecareplanning@fraserhealth.ca**

Six triggers for initiating discussion about end-of-life issues:[7, 10]

- The patient or family asks – or 'opens the door' – for end-of-life information and/or interventions (elicited or spontaneous, verbal or non-verbal).
- Psychological and/or social or spiritual distress or suffering.
- Pain requiring higher doses of analgesic medications.
- Dysphagia requiring feeding tube.
- Dyspnea or symptoms of hypoventilation, a forced vital capacity of 50% or less.
- Loss of function in two body regions (regions include bulbar, arms and legs).

Recommendation 4 Symptom Treatment

Swallowing and Nutrition:

- Dysphagia is caused by disturbed motility of the tongue, pharynx and esophagus.[2, 9]
- Nutritional consult and swallowing assessment.[1, 4, 5]
- Patients with bulbar symptoms early in disease may benefit from enteral feeding (PEG tube).[3, 5, 7, 8, 11] Consideration for PEG placement becomes critical with loss of greater than 10% of pre-diagnostic body weight[1, 2, 5, 6, 8, 9], for symptomatic dysphagia and before forced vital capacity falls below 50% of predicted.[1-3, 6, 8, 10, 12] Aspiration may occur with overfeeding through a PEG tube.[2, 5, 6, 9]
- Nutritional and fluid support in the final stages may not be indicated.[4, 5, 8]
- Patients and families should be informed that decreasing appetite and interest in food is a natural part of chronic disease.[4]

Recommendation 4 Symptom Treatment continued...

Choking:

- The fear of 'choking to death' is on the mind of many patients living with ALS. Yet, a study of 171 patients who died with ALS found that no patient choked to death.[2, 4, 11] This is helpful information to share with patients and families to relieve unwarranted fears of choking.[2, 11]
- Educate patient and family about techniques to minimize choking, such as changing diet consistency (thickened fluids and decreasing crunchy or chewy foods), positioning and swallowing techniques (Access Nutrition, Speech Language Pathologist or Occupational Therapist consultation).[2, 4, 8, 9]
- Teach family and caregivers that a calm reassuring presence assists in management of this symptom.[2, 9]
- Consider having a suction device available.[4, 10] Suctioning has been found to be most effective in patients who have a tracheostomy.[2, 6, 9]
- Manually assisted coughing techniques and mechanical insufflation-exsufflation devices are effective in clearing excess mucous.[1, 4, 6, 8, 12]
- Consider lorazepam sublingual or subcutaneous injection available for times of distress.[11]

Sialorrhea (drooling):

- Saliva production can be decreased in ALS patients but it becomes a problem when swallowing mechanisms are affected.[1, 5, 12]
- It is important to differentiate between sialorrhea and thick mucous production as treatments vary between the two causes.[1, 8, 12]
- It is more difficult to initiate a swallow with dry mouth, as saliva acts as an important part of the oral prep phase of the swallow. If dry mouth occurs, medications can be given with a puree food item, such as apple sauce or pudding.
- The following medications can be used to reduce secretions, but be cautious that the drying effect is not beyond normal or comfortable moisture levels. The mouth should never be so dry that swallowing cannot be easily initiated. The intramuscular route should be avoided in ALS patients due to muscle wasting.[4]

Anticholinergic affects of drugs producing a drying effect in the mouth:

- Glycopyrrolate[4, 6, 9] 0.6 to 1 mg t.i.d. PO[1, 12] OR 0.1 to 0.2 mg S.C. t.i.d.[6, 9]
- Transdermal scopolamine patch[1, 5] q72 hours.[4]
  - Transdermal patch absorption can be inconsistent in patients with excessive perspiration.[1, 8, 12]

Recommendation 4 Symptom Treatment continued...

- Buscopan 10 to 20 mg PO or per rectum q4h to q6h.[4, 6, 9]
- Amitriptyline[8, 12] 10 to 150 mg PO daily.[4]
- Atropine[6, 9] 0.4 to 0.6 mg PO OR S.C. b.i.d. or t.i.d. (4) OR 0.25 to 0.75 mg daily.[8]
- When oral medications fail to control sailorrhea, botulinum toxin A.[8, 12] 7.5 to 20 units injected into the parotid gland (and submandibular gland if the parotid glands alone are not beneficial) is an alternative.[8]
- Thick mucous production associated with sialorrhea can be treated with propranolol or metoprolol.[4] Guaifenesin is also useful.[1, 5]

Dyspnea

- Provide information that shortness of breath is a symptom that can be managed by medications and other techniques[2, 9, 11] (see *Fraser Health Hospice Palliative Care Symptom Guideline for Dyspnea*).
- Patient and family should have early education that medications are available when needed for respiratory insufficiency or pain management.
- Assess to determine if assisted ventilation through BiPap or other non-invasive ventilation device would be beneficial.[1, 2, 5-11] Non-invasive ventilation devices are often instituted when forced vital capacity is less than 50%.[8] Mechanical ventilation is seldom appropriate in end stage disease. Some clients are well established on BiPap, which is non-invasive.
- Loss of bulbar muscle tone and difficulty clearing secretions reduce tolerance of non-invasive devices and may define the limit of their use.[12]
- Position so that respiratory movement is facilitated.[9]
- Use fresh air moving past the face, relaxation and reassurance to relieve shortness of breath.[4]
- Pace activities that provoke increased breathlessness.[5]
- Use oxygen for hypoxia only.[9]
- **Treat reversible causes.**[9]
  - Initiate opioids when dyspnea is uncomfortable.
  - For pharmacologic management (*see Fraser Health Hospice Palliative Care Symptom Guideline for Dyspnea*).

Recommendation 4 Symptom Treatment continued...

Pain

- Usually musculoskeletal in origin, secondary to immobility.[1, 4-6]
- Severe neck pain, secondary to "floppy head" syndrome (weakness in neck extensors and flexors), can be relieved with the use of a hard Philadelphia or Freeman type collar.[1, 5]
- Utilize relaxation and diversion to help minimize pain.[10]
- Regular NSAIDS are useful when pain is mild.[1, 4-6, 10-12] They are particularly useful when active inflammation is present (as in arthritis or tenosynovitis).[1, 5]
- Acetaminophen 1000 mg PO q6h can be used with NSAIDS or alone.[5, 6]
- Many patients require regularly scheduled opioids as disease progresses, titrated to point of comfort.[1]
- Hydroxyzine 50 mg q6h potentiates analgesic effect of the narcotic.[1, 4, 8] It is a direct skeletal muscle relaxant that is not a cortical depressant.[5]
- For pain with a neuropathic component antidepressants and antiepileptics (gabapentin) are sometimes helpful[1, 4, 12] (*see Fraser Health Hospice Palliative Care Symptom Guidelines for Pain Management).*

Spasticity

- Regular stretching is a key to managing spasticity[1, 5, 8] especially for the gastrocnemius muscles.[1]
- Use gentle physiotherapy, positioning, splints, etc., as appropriate.[1, 4-6, 8]
- Range of motion exercise and massage has been shown to decrease spasticity and pain.[4, 7, 8, 10]
- Use baclofen[6, 9] 5 to 10 mg b.i.d. to t.i.d. up to 20 mg t.i.d.[1] OR 10 to 80 mg daily.[6, 9]
- Tizanidine (can be added if required) 4 to 8 mg t.i.d. to q.i.d.[1] OR 6 to 24 mg daily.[6, 9]

Dysarthria/Communication

- Educate patients and family early about management of speech and communication issues that may arise at the end of life.[4, 6, 8, 10]
- Refer to an Occupational Therapist or Speech Language Pathologist for communication tools.

Recommendation 4 Symptom Treatment continued...

Depression

Reactive depression is common in ALS patients.[1, 9] Up to 75% of ALS patients self report depressive symptoms.[6, 8, 9]

- For treatment of Depression see the *Fraser Health Hospice Palliative Care Symptom Guidelines for Depression*.
- When selecting an anti-depressant consider that amitriptyline is also useful in controlling pseudobulbar effect, drooling and sleep disturbances.[4, 6, 9]

References

Information was compiled using the CINAHL, Medline (1996 to April 2006) and Cochrane DSR, ACP Journal Club, DARE and CCTR databases, limiting to reviews / systematic reviews, clinical trials, case studies and guidelines / protocols using ALS terms in conjunction with palliative / hospice / end of life / dying. Palliative care textbooks mentioned in generated articles were hand searched. Articles not written in English were excluded.

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