



Hospice of the Twin Cities

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“It takes courage for people to listen to their own goodness and act on it.”
~ Pablo Casals ~

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Symptom Management of the Patient with ALS

Symptom Management of the Patient with Amyotrophic Lateral Sclerosis, A Guide for Hospice Nurses, by Gail Houseman, APRN which is published in the *Journal of Hospice and Palliative Nursing*, August/September 2008, Volume 10, Number 4, pp. 207-215 is an excellent article. I would like to share many of the symptom management techniques suggested in this article (the “Reader’s Digest” version).

What is Amyotrophic Lateral Sclerosis (ALS)?

It is a progressive, neuromuscular disorder that involves the degeneration of both the upper and lower motor neurons leading to eventual wasting and atrophy of all voluntary muscles, including muscles that are used for respiration (p.207). Respiratory failure is the usual cause of death for those with ALS. About 1-2 people in 100,000 are affected by this disease and the usual life expectancy, after diagnosis, is 2-5 years. There is no predictable progression of this disease (p. 207). The majority of patients diagnosed with ALS are men, and the onset is usually between 20-90 years of age. Research shows that more than 50% of those with ALS suffer some cognitive impairment, usually problems with judgment.

What are the most common onset of symptoms?

These symptoms vary, just as the disease progression varies from patient to patient. Unexplained weakness in a limb is a common symptom. Others can occur in swallowing and speaking muscles. There are others who develop symptoms in the respiratory muscles, and it is these patients who have the poorest prognosis. At some point in the disease process, nearly everyone develops respiratory difficulties leading to respiratory failure. About 90-95% of ALS patients choose not to have invasive mechanical ventilation when respiratory failure occurs (p.208).

Is there a consensus amongst physicians treating those with ALS when it comes to symptom management?

Sadly, there is not a consensus when it comes to treating the symptoms one with ALS might exhibit. Therefore, it is important to note that the recommendations that follow are the sole recommendations of the writer of the article,

Gail Houseman, APRN, based on her participation in a multidisciplinary team at a National ALS Association-certified ALS center in Philadelphia, PA. (p.208).

SYMPTOM MANAGEMENT

Respiratory Insufficiency

The following may be exhibited as signs and symptoms of respiratory insufficiency: SOB, dyspnea on exertion, orthopnea, waking up frequently/poor sleep, and morning headaches. The primary treatment for respiratory insufficiency in the ALS patient is BiPAP (Bilevel positive airway pressure). This air pressure is applied using a mask and has two levels of air pressure. A greater pressure is exerted on inhalation and a lesser pressure is exerted on expiration. The pressure can be adjusted to meet the patient’s need. There are generally three types of masks to choose from. The first is a mask that covers the nose, the second covers the nose and mouth, and the third is nasal pillows that rest in the nostrils. The last mask is good for patients who have skin breakdown from either of the first two examples. Oxygen is contraindicated for ALS patients unless it is used in conjunction with the BiPAP. With respiratory muscle weakness, if oxygen is given on its own, the ALS patient could have a buildup of carbon dioxide resulting in respiratory failure (p. 209).

Fatigue and Mobility Issues

Fatigue occurs secondary to muscle weakness and loss. When the ALS patient also has respiratory issues, they use a lot of their energy just breathing. Conservation of energy is the key for anyone with a chronic illness, and of the highest priority for those with ALS. Patients must learn how to balance activity with rest, and this is a difficult task for those who have been active throughout their lives. It is important that the caregivers are continuously assessing the ALS patient, adding medical equipment, as necessary, and performing ROM (range of motion) as part of the patient’s exercise regimen. Contractures can occur in any of the ALS patient’s joints and can be quite painful.

Pain

Estimates are that as many as 2/3 of ALS patients experience pain as part of their disease symptoms. Muscle spasms can be relieved by

Specific Guidelines for determining Prognosis: Amyotrophic Lateral Sclerosis (ALS)

Must fulfill 1, 2, or 3

1. **The patient must demonstrate critically impaired breathing capacity as evidenced by all of the following occurring within the twelve months preceding initial hospice certification.**
 - A. Vital capacity (VC) less than 30% of normal.
 - B. Significant dyspnea at rest.
 - C. Requiring supplemental oxygen at rest.
 - D. Patient declines artificial ventilation.
- 2.1. **Patient must demonstrate rapid progression of ALS as evidenced by all of the following occurring within the twelve months preceding initial hospice certification.**
 - A. Progression from independent ambulation to wheelchair or 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 or all activities of daily living (ADLs) to needing major assistance by caregiver in all ADLs.
- 2.2. **Patient must demonstrate critical nutritional impairment as evidenced by all of the following occurring within the twelve months preceding initial hospice certification.**
 - A. Oral intake of nutrients and fluids insufficient to sustain life.
 - B. Continuing weight loss;
 - C. Dehydration or hypovolemia;
 - D. Absence of artificial feeding methods.
3. **Patient must demonstrate both rapid progression of ALS (2.1 above) and life-threatening complications as evidenced by all of the following characteristics occurring within the twelve months preceding initial hospice certification.**
 - A. Recurrent aspiration pneumonia (with or without tube feedings);
 - B. Upper urinary tract infection, e.g., Pyelo-nephritis;
 - C. Sepsis;
 - D. Recurrent fever after antibiotic therapy.

Symptom Management for Patients with ALS, cont.

antispasmodics (Baclofen and Zanaflex). The following can also be tried for muscle spasms: Heat or cold and providing slow, gentle stretches and passive ROM. For charley horse kinds of cramps quinine (tonic) water can be tried, as well as trying Neurontin or other anticonvulsants. Some ALS patients complain of burning or numbness in their lower extremities and it is recommended to try elevating the patient's legs or trying Neurontin (p.209).

Dysarthria

All ALS patients should be evaluated by a speech language pathologist and if possible have an appropriate augmentative communication device in place before admission to hospice. The following are examples of appropriate devices: Communication card or board, writing board, text to speech devices, or eye gaze systems (pp. 209-210).

Dysphagia

This is a significant problem for those with ALS. Some patients have trouble chewing and swallowing, but can speak intelligibly. Some have feeding tubes placed to aid with nutrition. Thin liquids are a problem, as are foods with more than one consistency (i.e. soup with vegetables in it). Smaller, more frequent meals are recommended, using foods higher in calories, as well as supplements such as Ensure or Boost, and try to avoid dry foods. Aspiration is a common problem. Encourage the patient to eat slowly concentrating on chewing and swallowing, tucking his/her chin, sitting up while eating and after for a period of time, and taking medications crushed in pudding, yogurt, or applesauce.

Problems With Secretions

Droling (sialorrhea) can be embarrassing for the ALS patient. This occurs secondary to oral-pharyngeal muscles weakening and can be treated by anticholinergic medications such as Transderm Scop, Levsin, or Robinol. Anticholinergic antidepressants can also be prescribed, such as Elavil and Nortriptyline. If these medications are ineffective some patients try Botox injections into the parotid and submandibular glands. Thick mucus can accumulate in the back of the throat due to weakness in the throat muscles and a decreased ability to swallow. This often causes a fear of choking on their own fluids. Sometimes ALS patients must sleep sitting up to prevent choking on their own mucus. There are medications that can be used to thin secretions including quafenesin and Mucinex, Albuterol or Mucomyst nebs, inhaled nasal steroids (Flonase and Nasocort), and oral-pharyngeal suction may be helpful to stimulate a cough and remove mucus. There is also a device that can be used called insufflatorexufflator or Cough Assist (pp. 210-211).

Involuntary Emotion Expression Disorder

This is an upper motor neuron symptom which causes the patient to have difficulty controlling emotions, characterized by excessive and inappropriate laughing or crying. Antidepressants are usually effective in helping to control this symptom (p. 211).

Depression

Antidepressants may be helpful if the symptoms warrant their use. Supportive therapies and interventions may also provide relief (p. 211).

Insomnia

Insomnia may be caused by one of several factors: respiratory difficulties due to the patient lying in a supine position; depression; anxiety; pain; spasms; or cramps. The cause must be determined before deciding on the appropriate treatment (p. 211).

Constipation

This symptom is usually caused by changes in diet and mobility. If the patient is on pain medications, they will also affect constipation (p. 211).

Other Symptoms: Patients with ALS may also experience other symptoms as they progress through their illness, including urinary urgency and frequency, not related to urinary tract infection or enlarged prostate; laryngospasm; jaw clenching and tightening, including grinding teeth at night; cognitive issues (3-15% of all ALS patients meet the criteria for frontal temporal dementia (FTD); quality of life and spiritual concerns, and the terminal phase of their disease (p.211-212).

For more insight into this very complex disease and managing it's symptoms, please read this article in its entirety.



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Mission Statement

Hospice of the Twin Cities' mission is to enhance the quality of the lives of our patients and their families by providing respectful care based on maintaining dignity, alleviating physical, psychosocial, and spiritual suffering, advocating for fundamental rights, and affirming the sacred value of life.