What is ALS?

Introduction

Amyotrophic lateral sclerosis (ALS or Lou Gehrig's disease) is a progressive, incurable neurological disease that affects the nerve cells responsible for controlling voluntary muscles. The nerve cells that become affected take away the muscles ability to function and, as a result, the muscles gradually weaken, waste away, and twitch (fasciculation). Although a person diagnosed with ALS will eventually not be able to move without assistance, he or she will still be able to control their extraocular (eye) muscles and rectal/urinary sphincters. An article in March 2006, showed evidence that almost a third of the people diagnosed with ALS may demonstrate some cognitive impairment or dementia however; the disease does not usually affect a person's thinking or cognitive abilities.

What is the Prognosis?

Individuals will experience progressive weakness with moving, swallowing, speaking and breathing. ALS also progresses with weakness, wasting and paralysis of the muscles of the limbs and trunk as well as those that control speech, swallowing and breathing.

As the disease advances, choking spells become common when the person attempts to eat or drink, due to jaw weakness and fatigue. These episodes can be very frightening for the patient and caregivers. Nutritional support can be provided via a feeding tube and this can provide another route for medication administration. Recommendations for placement of a feeding tube are made soon after the onset of dysphagia and the decision about whether or not to insert the feeding tube is based on prolongation and quality of life.

When muscles in the diaphragm and chest wall fail, individuals lose the ability to breathe without the support of mechanical ventilation. Even though a respirator can ease problems with breathing and prolong survival, it does not affect the progression of the disease. Most people with ALS die of respiratory failure or pneumonia, not the disease itself.

Palliative Care

Pharmacological and non-pharmacological interventions should focus on symptomatic relief, prevention of complications, maintenance of maximum function and optimal quality of life.

Pharmacological treatment of ALS includes Riluzole (Rilutek©), the only FDA approved medication for the treatment of ALS. Riluzole may prolong life by a few

months in ALS patients by slowing down the progression of the disease. Sialorrhea, an excessive secretion of saliva, is a prominent symptom in people with ALS. Pharmacological interventions include Robinal (Glycopyrrolate), Cogentin, Scopolamine patch, Atropine or Elavil (Amitriptyline).

Pseudobulbular effect or pathological laughing or crying may be seen in up to 50% of people with ALS. Pharmacological treatment includes Elavil (amitriptyline) or Fluvoxamine (Luvox).

Pain may be present from stiff joints, muscle cramps or a preexisting condition. Refer to your pain assessment and guidelines for pain management and bowel regimen treatments.

Substantial physical caregiving is required for the person diagnosed with ALS. Common equipment needs may include: electronic wheelchair, letter & picture board for communication, BiPap machine, feeding tube equipment.

The options for respiratory support should be discussed before the development of respiratory muscle weakness. Involvement of respiratory muscles and Upper Respiratory Infection (URI) can lead to sudden decline and death. Signs and symptoms of respiratory muscle weakness include dyspnea with exertion, dizziness, inability to lie flat without shortness of breath, anxiety, morning headaches, weight loss (may be due to decreased intake) and extreme fatigue. Discussions concerning the choice of mechanical ventilation should take place between patient, caregivers and healthcare professionals before a crisis situation occurs.

Aggressive psychological and spiritual support is needed to assist patients with ALS and their families as these patients often experience fear, anxiety and depression. Support groups including online chat groups are an excellent resource for patients and their caregivers. To assist patients and caregivers in finding a support group, contact your local ALS organization or Muscular Dystrophy Association (MDA) office.

When is the Right Time for Hospice?

According to the ALS organization, the majority of people diagnosed with ALS die from respiratory failure, usually within 3 to 5 years from the onset of symptoms. Approximately twenty percent of people with ALS will live five years or more, up to ten percent will survive more than ten years and five percent will live 20 years from time of diagnosis with ALS. Hospice care is an important part of the care a person with ALS receives, but when is the right time for hospice?

When evaluating a potential hospice admission with a person diagnosed with ALS, hospices need to consider many issues including the artificial hydration, nutrition and mechanical ventilation which can significantly prolong the life expectancy of the ALS patient. Goals of care should be carefully discussed and agreed upon by the patient, caregiver and clinicians at the time of admission to hospice.

Documentation

Document the following signs and symptoms:

- Progression of disease in linear fashion
- Independent ambulation to wheelchair or bedbound
- Normal to barely or unintelligible speech
- Normal to blenderized diet
- Independence in ADL's to requiring major assist
- Multiple clinical parameters required to judge the progression of disease
- Rate of progression is important in predicting prognosis.

Critical Factors in Determining Prognosis

Ability to breathe as demonstrated by:

- Impaired ventilatory capacity
- Vital capacity less than 30% of predicted
- Significant dyspnea at rest
- Supplemental oxygen required at rest
- Critical impaired respiratory status can be measured with a Forced Vital Capacity (FVC) less than 40% and two of the following signs and symptoms:
  - ✓ Dyspnea at rest
  - ✓ Orthopnea
  - ✓ The use of accessory muscles to breath
  - ✓ Paradoxical abdominal motion
  - ✓ Reduced speech/volume
  - ✓ Weak cough
  - ✓ Symptoms of sleep disorder breathing
  - ✓ Unexplained headache
  - ✓ Confusion
  - ✓ Anxiety
  - ✓ Nausea

If the patient cannot perform the Forced Vital Capacity test, documentation should reflect that at least three of the above signs and symptoms are present.

Ability to swallow as demonstrated by:

- Nutrition impairment: Nutritional status is defined if dysphagia is present with weight loss of 5% of body weight with or without a feeding tube in place.

- Oral intake insufficient to sustain weight
- Weight loss
- Dehydration or hypovolemia

The following factors are further indications of decreased survival time. If present, document the following:

- Recurrent aspiration
- Pneumonia
- Decubitus ulcers, multiple
- Upper urinary tract infection
- Sepsis
- Fever recurrent after antibiotics

Documentation Tips

Remember that ALS tends to progress in a linear fashion and that the history of rate of progression is critical in determining/predicting prognosis. Document all signs and symptoms including onset and change over time. Include information relating the psychosocial and spiritual needs and interventions which impact the overall quality of life.

References

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