FAST FACTS AND CONCEPTS #73
MANAGEMENT OF RESPIRATORY FAILURE IN ALS

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Background Respiratory failure is the most common cause of death from amyotrophic lateral sclerosis (ALS). Except for rare patients who present with respiratory failure, respiratory muscle weakness develops insidiously during the course of the disease. This allows most patients, families, and clinicians time to carefully discuss the options for ventilatory support before such decisions needs to be made. *Options for ventilatory support should be discussed long before the development of respiratory insufficiency.* In addition, patients should be encouraged to identify a power of attorney for health care/ healthcare proxy and to discuss their preferences about ventilatory support early in the course of the illness.

Assessment of Respiratory Insufficiency Early signs and symptoms of respiratory muscle weakness are subtle: dyspnea with mild exertion, supine dyspnea, insomnia, morning headache, reduced appetite, weight loss, dizziness, depression, anxiety and marked fatigue. There are no standard protocols to detect early respiratory failure. The following tests may be helpful:

- *Maximal Inspiratory Pressure* (MIP) has been reported to be the single most sensitive test.
- Measuring the change from erect to supine sitting *Vital Capacity* (VC) can detect early diaphragmatic weakness and can be used to monitor declining respiratory function. Diaphragm weakness is suggested by a greater than 25% fall in VC on assuming a supine position. A normal supine VC makes significant inspiratory muscle weakness unlikely. A VC less than 1L or less than 30% of predicted value indicates significant risk of respiratory failure and death.
- *Nocturnal oximetry* is useful in evaluating nocturnal hypoventilation; a full polysomnogram can be an alternative test.

Choices for Respiratory Management

- *Bi-level Positive Airway Pressure* (BiPAP or bilevel support), a type of non-invasive ventilation, can improve symptoms of hypoventilation, quality of life, and survival by several months. Inability to safely clear secretions, which is common in those with bulbar involvement, is a relative contraindication to bilevel support. In fact, inability to clear secretions secondary to progressive ALS is often used as a clinical sign to indicate that bilevel support should be discontinued. The use of oxygen at night without ventilatory assistance may not be sufficient for many patients with advanced ALS with life prolonging goals of care, as oxygen desaturation and chronic hypercapnia may lead to suppression of respiratory drive during sleep. During the day bi-level support can be used to alleviate respiratory muscle fatigue. The use of 'intermittent positive pressure' breathing machines has been reported to expand the lungs and reduce atelectasis.

- *Full-time invasive ventilatory support* is considered when non-invasive ventilation is no longer effective or tolerable. Treating reversible problems (e.g. acute respiratory infection) is important to avoid the need for invasive long-term ventilation with a tracheostomy. Most patients who select a ventilator become completely dependent on it and are unable to communicate verbally. Many patients will choose not to be ventilated if comfort is assured and adequate advance care planning occurs.

Supportive Care Regardless of assisted ventilatory choices, compassionate and effective palliative care must be implemented. A calm environment, the reassuring presence of relatives, trunk elevation and chest physiotherapy may all provide relief. The sensation of shortness of breathing can be reduced by morphine (2.5-10 mg by mouth or 1-2 mg IV/subcutaneous every 1-4 hours). Titration of the morphine dose against the clinical effect almost never leads to a life threatening respiratory depression (see *Fast Fact* #27). Anxiety due to respiratory insufficiency can be treated with lorazepam (0.5-1.0 mg sublingually). When co-morbid medical complications develop, or patient determined quality of life deteriorates, ventilator support can be withdrawn and symptom control provided to allow for a comfortable death. See *Fast Facts* # 33, 34, 35 about ventilator withdrawal. See *Fast Facts* # 299, 300, and 301 regarding management of sialorrhea and other common symptoms in ALS.

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