



## The Value of Pulmonary Function Testing

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Amyotrophic lateral sclerosis (ALS) eventually results in progressive respiratory muscle weakness and trouble moving air in and out of the lungs, leading to breathing failure unless respiratory symptoms are treated. When breathing failure is prevented or treated and complications are avoided, ALS itself is not fatal. People with ALS have the right to know about respiratory care options, methods available to continue breathing and palliative care alternatives if resuscitation is not desired.

Early detection of respiratory impairment is essential for timely interventions in planning and implementing appropriate care, reducing the risk of emergency hospitalizations. Thus, the key to a successful treatment outcome is careful monitoring of respiratory symptoms that includes periodic pulmonary function testing.

### **Signs of Respiratory Muscle Weakness**

People with ALS frequently deny trouble breathing. Although some people appear to have shortness of breath on exertion or rest, visible signs of breathing disturbance may be overlooked unless respiratory impairment is advanced. Despite the

absence of noticeable signs and denial of trouble breathing, telltale signs of respiratory muscle weakness include inability to take a deep breath, especially while lying flat, a weak cough and low voice volume.

However, clinical signs alone cannot always determine pulmonary function accurately. Patients, therefore, should see their physician who may order pulmonary function testing to monitor their symptoms.

### **Value of Pulmonary Function Testing**

Pulmonary Function Tests (PFTs) reveal the degree of respiratory muscle involvement, the progression of symptoms, early recognition of reversible complications, pending respiratory failure and the need for appropriate therapy as desired by the patient. PFTs may be more sensitive than a history and physical examination in accurate recognition of pulmonary function. PFTs can be done simply and non-invasively, and if prescribed by a physician, can be done at home by a respiratory care professional or even by the patient.

### **Types of Pulmonary Function Tests**

**Spirometry** or measurement of the air capacity of the lungs is commonly used in people with ALS and includes a variety of tests. The **Forced Vital Capacity** measures the total amount of air that can be forcefully exhaled out of the lungs after maximal inhalation. The patient wears a nose clip so no air escapes from the nose. The patient is instructed to breathe in as deeply as possible through the mouth and hold the breath. The lips are firmly placed around the mouthpiece that is attached to the hose connected to a spirometer, a computerized or hand-held measuring apparatus. Then the patient is instructed to blow all the air out as quickly and forcefully as possible.

If the patient is unable to keep a tight seal around the mouthpiece, then an oral / nasal mask should be used. For the most accurate assessment, the test should be done both in sitting and lying down positions. If the vital capacity is lower when lying down than sitting upright, this usually indicates diaphragm muscle weakness.

Accurate spirometry testing depends on the patient's cooperation and effort.

Because a full stomach limits lung expansion that may alter test results, the patient should not eat one hour beforehand. Fatigue may also lower test results, thus, tiring activities should be postponed prior to the testing. The patient should also wear loose clothing.

**The Forced Vital Capacity values** are compared to the "predicted normal" values of persons who are similar to the patient in age, height and sex. Results are usually indicated as a percentage of the predicted value. A value of 85% or higher of the predicted is considered normal. A FVC that is 50 - 60% of the predicted normal reveals a moderate abnormality, signaling the time to be evaluated for assisted ventilation using a mechanical device if desired by the patient. A 25% to 30% FVC or lower indicates that breathing failure would be expected unless appropriate treatment is given.

The **Peak Expiratory Flow** measures the cough force or the ability to forcefully exhale and is a test that can be done easily at home. The maximal airflow during forced exhalation, after full inspiration, can be measured with a small portable peak flow meter. This is an inexpensive device that can be obtained at a local pharmacy or medical supplier.

Other tests include the **Maximum Inspiratory Pressure (MIP)** and the **Maximum Expiratory Pressure (MEP)**. They test

the strength of the inspiratory and expiratory muscles. The MIP and MEP are measured with a manometer, a pressure-monitoring device. The MIP is the highest pressure that can be generated during inspiration and the MEP is the highest pressure that can be generated during an expiration, through a device with a closed airway.

**Pulse Oximetry** is non-invasive electronic monitoring of the oxygen saturation in the blood. A visiting nurse or respiratory care practitioner can do this conveniently and quickly at home. The test is done painlessly without needles by clipping a photoelectric device over a finger for spot-checking or continuous monitoring. Pulse oximetry can detect low oxygenation in the blood before respiratory symptoms are noticeable. Normal oxygen saturation is 95% or higher.

**Nocturnal Pulse Oximetry** is sometimes ordered to monitor oxygen saturation during sleep at night to identify hypo-ventilation, a slow, shallow breathing pattern. If the oxygen saturation is below 90%, nasal ventilation might be considered to assist breathing. Pulse oximetry is the best alternative to expensive, painful arterial blood gases, requiring a needle puncture, which may not reveal abnormal oxygen and carbon dioxide levels in the blood until breathing problems are advanced.

People with ALS should know why pulmonary tests are performed and the meaning of

their test findings to keep informed of their respiratory status and when treatment may be needed. As never before, options are available to assist breathing and treat symptoms, while reducing the risk of complications.

Those who do not have excessive oral secretions may be the best candidates for trying a ventilator that administers positive pressure air through a nasal mask, nasal conduits or a mouthpiece. Use of nasal / oral ventilation with a bi-level pressure or a volume-cycled ventilator prolongs survival in selected individuals. This is an alternative to long-term use of tracheostomy ventilation. Obtaining accurate and sufficient information alleviates the fear of what lies ahead and helps prevent hasty decisions. It is better to plan treatment well ahead than to be one minute late.

#### References:

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