Tests for Breathing Problems If You Have a Neuromuscular Condition

Prepared by International Ventilator Users Network (IVUN) www.ventusers.org
Reviewed by Nicholas S. Hill, MD, Tufts-New England Medical Center, Boston, Massachusetts

If you have a neuromuscular condition such as post-polio syndrome, ALS, or Duchenne muscular dystrophy, you may not realize that your breathing muscles are weak and can become weaker. You may have difficulty breathing in deeply enough to fully expand your lungs or coughing strongly enough to clear mucus from your lungs.

It is essential to have periodic measurement of your respiratory muscle function and strength, and your oxygen and carbon dioxide levels to determine whether you may need to use a breathing machine (bilevel device or ventilator) during the night and/or need help with coughing. If you are a polio survivor, measurements of forced vital capacity (the maximum amount of air one can exhale) should be taken BOTH when you are sitting upright and lying down (in the supine position).

Even after you begin using assisted ventilation (a bilevel device or ventilator) it is critical that you periodically have your breathing monitored to prevent respiratory complications in the future and to improve your quality of life and survival. The recommendations below were written for individuals with Duchenne muscular dystrophy, but they are also appropriate for people with ALS and post-polio syndrome.

Routine Evaluation of Respiratory Function

- Objective evaluation at each clinic visit should include: oxyhemoglobin saturation by pulse oximetry, spirometric measurements of FVC, FEV_i, and maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures, and peak cough flow.
- Awake carbon dioxide tension should be evaluated at least annually in conjunction with spirometry. Where available, end-tidal capnography can serve the purpose of monitoring CO2 levels. The need for arterial blood gas analysis will depend on your doctor's recommendation. If capnography is not available, then a venous or capillary blood sample can be obtained to assess for the presence of alveolar hypoventilation (too little breathing causing CO2 to increase).
- Additional measures of pulmonary function and gas exchange may be useful, including lung volumes and assisted peak cough flow.
- Careful evaluation of patients for evidence of other respiratory disorders, such as obstructive sleep apnea, oropharyngeal aspiration, gastroesophageal reflux, and asthma.
- Annual laboratory studies in patients requiring a wheelchair for ambulation should include a complete blood count, serum bicarbonate concentration, and a chest radiograph.

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Medical Journal Articles: Breathing and Sleep Problems in Neuromuscular Conditions

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