

## **Tests for Breathing Problems If You Have a Neuromuscular Condition**

Prepared by International Ventilator Users Network (IVUN) [www.ventusers.org](http://www.ventusers.org)

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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<sub>i</sub>, 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 CO<sub>2</sub> 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 CO<sub>2</sub> 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.

Finder JD, Birnkrant D, Carl J, et al. Respiratory care of the patient with Duchenne muscular dystrophy. *Am J Respir Crit Care Med.* 2004;170: 456-65. Official Journal of the American Thoracic Society. ©American Thoracic Society. Reprinted with permission.

## **Medical Journal Articles: Breathing and Sleep Problems in Neuromuscular Conditions**

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### **General**

Simonds AK. Recent advances in respiratory care for neuromuscular disease. *Chest* 2006; 130:1879-1886

### **ALS**

Bach JR. Bilevel pressure vs. volume ventilators for amyotrophic lateral sclerosis. *Chest* 2006; 130: 1949

Bourke SC, Tomlinson M, Williams T, et al. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurology* 2006; 5:140-147

Lechtzin N, Wiener CM, Clawson L, et al. Use of noninvasive ventilation in patients with amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2004; 5(1): 9-15

Servera E, Sancho J. Appropriate management of respiratory problems is of utmost importance in the treatment of patients with amyotrophic lateral sclerosis. *Chest* 2005; 127:1879-1882

### **Cough**

Chatwin M, Ross E, Hart N, et al. Cough augmentation with mechanical insufflation/exsufflation in patients with neuromuscular weakness. *Eur Respir J*, 2003; 21:502-508

Lechtzin N, Shade D, Clawson L, Wiener CM. Supramaximal inflation improves lung compliance in subjects with amyotrophic lateral sclerosis. *Chest* 2006; 129:1322-1329

### **Duchenne muscular dystrophy**

Finder JD, Birnkrant D, Farber C, et al. Respiratory care of the patient with Duchenne muscular dystrophy; ATS consensus statement. *Am J Respir Crit Care Med* 2004; 170:456-465

Kohler M, Clarenbach CF, Böni L, et al. Quality of life, physical disability, and respiratory impairment in Duchenne muscular dystrophy. *Am J Respir Crit Care Med* 2005; 172:1032-1036

Toussaint M, Steens M, Wasteels G, Soudon P. Diurnal ventilation via mouthpiece: survival in end-stage Duchenne patients. *Eur Respir J* 2006; 28:549-555

### **Post-polio syndrome/Late effects of polio**

Bach J. Management of post-polio respiratory sequelae. *Ann NY Acad Sci* 1995; 25, 753: 96-102.

Bach J, Alba, A. Pulmonary dysfunction and sleep disordered breathing as post-polio sequelae: evaluation and management. *Orthopedics* 1991; 14:1329-1337

Gay, PC, Edmonds, LC (1995). Severe hypercapnia after low-flow oxygen therapy in patients with neuromuscular disease and diaphragmatic dysfunction. *Mayo Clin Proc* 1995; 70:327-330

Hsu, A, Staats, B. Postpolio sequelae and sleep-related disordered breathing. *Mayo Clin Proc* 1998; 73:216-224

Soliman M, Higgins S, El-Kabir DR, Davidson A, Williams, A, Howard, R. Non-invasive assessment of respiratory muscle strength in patients with previous poliomyelitis. *Respir Med* 2005; 99: 217-22