

## **Breathing Problems in Post-Polio**

**Articles from *Post-Polio Health* ([www.post-polio.org](http://www.post-polio.org)) and  
*Ventilator-Assisted Living* ([www.ventusers.org](http://www.ventusers.org))**

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*Post-Polio Health*, Vol. 20, No. 2, 2002

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### **Aging and Respiratory Function**

*Post-Polio Health*, Vol. 18, No. 1, 2004

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*Ventilator-Assisted Living*, Vol. 17, No. 1, 2003

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*Post-Polio Health*, Vol. 17, No. 2, 2001

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# Post-Polio Breathing and Sleep Problems Revisited

Judith R. Fischer, MSLS, Editor, *Ventilator-Assisted Living*, and Joan L. Headley, MS, Editor, *Post-Polio Health*

*“Post-Polio Breathing and Sleep Problems”* was published in the fall of 1995 (*Polio Network News*, Vol. 11, No. 4). As a result of the continual flow of phone calls and emails from polio survivors and family members about this life and death topic, Judith Fischer, editor of *Ventilator-Assisted Living* (our other quarterly newsletter), and I decided to revisit and revise the original article. Our goal is to educate and to clarify misinformation about breathing problems of polio survivors. —Joan L. Headley, Editor, *Post-Polio Health* (ventinfo@post-polio.org)

**N**ew breathing and sleep problems in aging polio survivors can be insidious and often go unrecognized by either polio survivors, their family members or their health care providers. Polio survivors may have weakened breathing muscles as a result of the initial damage by the poliovirus; the lungs themselves were not affected. Those who were in an iron lung during the acute phase should be aware of the potential for developing problems later in life and educate themselves in order to recognize important signs and symptoms which may indicate underventilation which may lead to respiratory failure. Even those who did not need ventilatory assistance during the acute phase may also be at risk for underventilation and should be aware of problems with breathing and sleep.

Underventilation (hypoventilation is the medical term) means that not enough air reaches the lungs to fully inflate them. The result may be too little oxygen and too much carbon dioxide (CO<sub>2</sub>) in the blood. Under-ventilation can be caused by one or more of the following: weakness of the inspiratory muscles (mainly the diaphragm and rib muscles) for breathing in, weakness of the expiratory muscles (the abdomen) for breathing out and producing an effective cough

to clear secretions, scoliosis (curvature of the spine), and sleep apnea.

Other factors contributing to a polio survivor's breathing problems are a history of smoking, obesity, undernutrition, and other lung diseases such as asthma, bronchitis and emphysema.

Vital capacity (VC) is the volume of air that can be expelled after taking a big breath and is a measure of how well the lungs inflate. VC normally decreases with age, but this decrease in VC is more serious in an aging polio survivor with weakened breathing muscles. Many polio survivors had impairment of their inspiratory muscles, and the normal changes due to aging may cause them to lose VC at a greater rate. Polio survivors may not experience symptoms of underventilation until their VC falls to 50% or less of predicted (normal).

## **Signs and symptoms of under-ventilation during sleep include:**

- ◆ inability to breathe when lying flat — the need to sleep sitting up (orthopnea)
- ◆ inability to fall asleep and/or to stay asleep (insomnia)
- ◆ anxiety about going to sleep
- ◆ restless fragmented sleep with frequent awakenings

Thanks to Lisa Krivickas, MD, Spaulding Rehabilitation Hospital, Boston, Massachusetts; E.A. Oppenheimer, MD (retired), Los Angeles, California; and Mark H. Sanders, MD, University of Pittsburgh Medical Center, Montefiore University Hospital, Pittsburgh, Pennsylvania, for their reviews of this article.

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- ◆ shallow breathing or pauses in breathing
- ◆ awakening from sleep with choking sensation
- ◆ nightmares, night sweats, bedwetting or need to urinate frequently
- ◆ excessive daytime sleepiness
- ◆ morning headaches
- ◆ worsening mental status and impaired memory, concentration and cognition

**Other symptoms may include:**

- ◆ shortness of breath on exertion
- ◆ fatigue or exhaustion from normal activities
- ◆ claustrophobia and/or feeling that the air in the room is somehow bad
- ◆ general anxiety
- ◆ difficulty in speaking for more than a short time
- ◆ low voice/volume speech with fewer words per breath
- ◆ use of accessory muscles, such as neck muscles, to breathe
- ◆ weak cough with increased respiratory infections and pneumonias.

Polio survivors experiencing one or more of the above signs and symptoms should seek a respiratory evaluation (simple and noninvasive pulmonary function tests) by a pulmonologist, preferably one experienced in neuromuscular disorders. Physicians are listed in the *Resource Directory for Ventilator-Assisted Living* ([www.post-polio.org/ivun/d.html](http://www.post-polio.org/ivun/d.html)).

**Pulmonary function tests should include the following measurements.** The values that indicate a warning sign for respiratory problems are in parentheses.

- ◆ VC — upright (<50%)
- ◆ VC — supine (a drop of >25% from upright to lying down)
- ◆ MIP — maximum inspiratory pressure (<60 cm H<sub>2</sub>O)
- ◆ MEP — maximum expiratory pressure (<60 cm H<sub>2</sub>O)
- ◆ peak expiratory cough flow (<300 L/min)
- ◆ end-tidal CO<sub>2</sub> (>45 mm Hg)
- ◆ overnight oximetry may be prescribed to detect episodes of oxygen desaturation (<88% during sleep).

Management of breathing and sleep problems can be achieved largely through the use of nocturnal noninvasive ventilation, commonly in the form of small, lightweight bilevel positive pressure units. The units have a long tube/circuit that attaches to a mask (nasal, facial or oral), nasal pillows or mouthpiece worn during sleep. Polio survivors may find themselves gradually extending periods of ventilator use, perhaps during a daytime nap. Some polio survivors may need to use a volume ventilator to guarantee delivery of a larger volume of air than a bilevel unit can provide. Noninvasive ventilation may eventually fail, and invasive tracheostomy positive pressure may be necessary.

Treating underventilation with oxygen therapy instead of assisted ventilation can lead to respiratory failure and death because supplemental oxygen

**Question:** *Why would my physician add oxygen to bilevel positive pressure ventilation, such as BiPAP®, if there is no underlying lung problem?*

**Answer:** It would be extremely worthwhile to request that PHI members who use a ventilator due to post-polio breathing problems ask that their physicians explain their oxygen advice so we can all benefit, or at the very least develop a list of justifications that can be reviewed. The question is simple and can be stated, “The use of oxygen for people with neuromuscular disease without any lung disease or pneumonia is a subject of considerable interest, and some controversy. Please explain why oxygen is advised as part of my post-polio treatment plan.” Send your physician’s comments to PHI (editor@post-polio.org).

A physician might advise adding oxygen to BiPAP® when there is no underlying lung problem, if the person is traveling by air, where the air has a lesser concentration of oxygen. Another possibility is that the bilevel device is not satisfactorily set up and not adjusted from time to time. If that is the case, the oximetry or ABG oxygen saturation might be below 95% and the physician might prescribe oxygen. The advised response to this, of course, would be to increase the IPAP (inspiratory positive airway pressure) to improve ventilation, and then to re-check the oxygen level until it stayed at 95% or better while using ventilation.

Reliable small portable oximeters, such as the Nonin Onyx® 9500, can now be purchased on the Internet without a doctor’s prescription. Having one at home to monitor assisted ventilation would allow minor adjustments of the IPAP as needed — if the treating physician agrees. Prices vary considerably on the Internet so search carefully. Most often an oximeter is not covered by health insurance.

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**Question:** *I am a polio survivor who uses a ventilator during the night. My physician wants me to undergo a colonoscopy, but I am worried about anesthesia for this procedure. Should I proceed?*

**Answer:** A colonoscopy is an important test that can be performed safely on polio survivors who use assisted ventilation. In your case, I suggest a pulmonary evaluation before the procedure to document your respiratory reserve and a candid discussion with whomever is performing the colonoscopy. Sedation is needed because this procedure is uncomfortable. Usually a gastroenterologist does the colonoscopy with a nurse giving the sedation and supposedly monitoring the patient.

The colonoscopy should be done in a hospital (not an outpatient surgery center) with an anesthesiologist in attendance and administering the sedation. The procedure can be in the hospital’s GI lab or in an operating room. The GI lab has adequate monitors; someone (the anesthesiologist) needs to watch you closely and to be careful with sedation. Afterward, you can be observed for respiratory problems in the regular recovery room and, if necessary, go to a “monitored” bed for an overnight stay — one night should be enough. You can bring and use your own home ventilator, just be sure this is discussed with the hospital staff ahead of time. I have received this question several times and after dispensing the above advice, I have not heard of any problems.

Selma Harrison Calmes, MD, Member of PHI’s Medical Advisory Committee  
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can blunt the function of the brain’s respiratory control center. However, polio survivors who use assisted ventilation and have additional medical problems such as COPD, pneumonia or heart problems, or who are undertaking long airplane flights, may benefit from oxygen therapy under careful supervision.

Polio survivors may also have sleep apnea contributing to underventilation. Sleep apnea, an interruption of breathing during sleep, can be obstructive,

central or mixed. Obstructive sleep apnea (OSA) is the most common form and is prevalent in the general population. The standard test for OSA is a sleep study; the standard treatment is the use of a continuous positive airway pressure (CPAP) unit with a nasal mask or nasal pillows during sleep. However, polio survivors with both weakened breathing muscles *and* sleep apnea should use bilevel positive pressure or volume ventilation, not CPAP. ●

*This article is adapted from a presentation at the Ninth International Noninvasive Ventilation Conference: "From the ICU to Home," Orlando, Florida, October 23-25, 2003.*

## **Aging and Respiratory Function**

**Anita Simonds, MD, FRCP, Royal Brompton Hospital,  
London, England**

Many factors need consideration in the transition to old age. These include the physiological effects of aging, the progression or late effects of the neuromuscular disease/condition, the effects of medication, the increased likelihood of surgery, and the quality of ventilatory support in care/nursing homes.



Social and psychological factors are also a major part of this transition. These include retirement, economic changes, loss of family ties, and the desire to retain one's autonomy and involvement in decision-making.

The effects of aging on respiratory function include:

- fall in lung volumes,
- decrease in area for oxygen exchange,
- reduced elasticity of chest wall,
- decrease in respiratory muscle strength,
- reduction in exercise level and deconditioning,
- osteoporosis, back pain, progression of scoliosis/kyphosis,
- cardiac problems that may add to breathlessness.

The increased likelihood of invasive procedures and surgery in later years necessitates careful risk/benefit assessment. Some ventilator users are told they are "not fit for surgery," but the risks of not carrying out the surgery must be weighed.

Many procedures and scans can be carried out safely under sedation, and individuals can use their own home ventilators, but liaison with the anesthetic team is vital. For pre-operative assessment, the surgical/anesthetic team must know the complete medical history, ventilatory equipment needs and care plan.

Some ventilator users may also need management and treatment for cancer/malignant disease. They need to ask questions: “What is the ideal treatment for my cancer — surgery, chemotherapy, radiation and/or hormone therapy? Is there any reason why I should not have treatment? If problems are anticipated, can we work around them?”

For ventilator users and people with chronic pulmonary diseases, pulmonary rehabilitation programs can improve exercise tolerance and increase quality of life.

Attributing new or progressive symptoms to “just getting older” without consideration of other possibilities is unwise. If you are a polio survivor, do not automatically ascribe new symptoms to the late effects of polio without eliminating other causes.

Aging brings changing goals and complex issues. There is often a lack of correlation between the views of the individual and those of the family and caregivers. Make sure your family, caregivers and health care team know your wishes — prepare advance directives and living wills.

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### **Observations by older ventilator users:**

- Coming off the ventilator in the morning is much harder.
- I am more breathless after a meal.
- I have side effects from my medication.
- My hearing loss prevents me from hearing the ventilator alarms.
- I worry about what will happen when I am on my own.
- I feel weaker.
- Because my legs are less strong, I am more unsteady on my feet.
- I tire more easily.
- I don't want to lose my independence.



## **What is Hypoventilation?**

**Robert A. Lebbly, MD**

The work of inspiring air, or breathing in, is performed primarily by the diaphragm although other muscles play a smaller role. With many neuromuscular diseases, these muscles become weak and less able to expand lungs and draw air in. Other factors making it harder to fully expand the lungs in some people, include lung and chest wall stiffness/inelasticity and scoliosis (spinal curvature).

Smaller breaths can result in less overall airflow or hypoventilation. If adequate airflow does not occur through the lungs, then carbon dioxide (CO<sub>2</sub>) is not effectively eliminated and may build up in the blood. Blood oxygen levels (O<sub>2</sub>) may also decrease due to the hypoventilation, high CO<sub>2</sub> levels, or other reasons.

The symptoms of hypoventilation may be subtle at first, especially in people with neuromuscular disease whose problems evolve very slowly allowing the body to adapt to the gradual changes. The earliest problems usually occur during sleep because the control of breathing changes with different sleep stages and because sleeping in the supine position may worsen breathing.

The signs and symptoms of nocturnal (sleep-related) hypoventilation include many that Bryna describes in her article:

- sleep initiation and sleep maintenance insomnia
- anxiety about going to sleep
- restless/fragmented sleep with frequent awakenings
- shallow breathing or pauses in breathing
- awakening from sleep with choking sensation
- nightmares, night sweats, bedwetting, or need to urinate frequently
- night-time or morning headaches
- excessive daytime sleepiness and need to nap during the day
- worsening mental status, impaired memory, concentration, cognition
- inability to lie flat during sleep/need to sleep sitting up (orthopnea).

Other symptoms of hypoventilation include:

- shortness of breath, breathlessness with minimal activity



- fatigue or exhaustion from normal activities
- claustrophobia and/or feeling that the air in the room is somehow bad
- anxiety
- difficulty in speaking for more than a short time
- quiet speech with fewer words per breath
- inability to lie flat (even while awake) due to shortness of breath.

Anyone experiencing a combination of these symptoms deserves a respiratory evaluation, preferably by a pulmonary physician experienced in neuromuscular diseases, sleep, and hypoventilation. Together, the physician and the individual can formulate a treatment plan for assisted ventilation.

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### **Additional thoughts from Peter C. Gay, MD**

**Associate Professor, Critical Care and Sleep Medicine, Mayo Graduate School of Medicine, Rochester, Minnesota**

**On weakness ...** "The vernacular defining post-polio syndrome includes non-specific weakness. Many physicians overlook the fact that polio survivors are describing respiratory problems."

**On symptoms ...** "The important thing to recognize is that the vast number of polio survivors do not necessarily have lung disease, per se. The problem usually is the inadequate way they move air in and out of their lungs.

"It is important to determine which symptoms are related to breathing, which are related to the heart, and which are related to deconditioning. Even Olympic athletes can become deconditioned."

**On abdominal paradox ...** "Many polio survivors have difficulty with breathing while lying. Physicians can assess this by observing the abdominal paradox which occurs when the abdomen inappropriately collapses inward as the chest expands during inhalation.

"Normally when one lies down and takes a deep breath, the diaphragm pushes down and the abdomen pushes outward. If the diaphragm is very weak and the chest muscles are doing most of the work, then the abdomen is going to paradoxically collapse. This is a very specific sign that a physician can recognize by placing a hand on the abdomen and asking the patient to

take a deep breath. If the abdomen collapses during inspiration, it is a sign of a weak diaphragm. During sleep, this is critical to understand."

**On deterioration of quality of sleep ...** "The sleeper that people get, the more they want to sleep, and the more they are tortured by the need to wake up to breathe."

**On sleep apnea ...** "The 'I won't breathe' form is central sleep apnea. The controller, during REM sleep particularly, stops firing impulses adequate to make one breathe. It is important to demonstrate the difference between these because different equipment is needed for each form.

"The 'I can't breathe because the upper airway is blocked' form is obstructive sleep apnea. It is most common in males and is due to being overweight and to the size of the neck and structure of the upper airway. In post-polio patients, the weakness of the muscles of the mouth can be such that, when lying on one's back, the muscles can block off and cause snoring."

**On nighttime assisted ventilation ...** "Breathing at night is like driving. You need power steering. You still want to drive, but it feels like you are driving a big Mack truck. You need a device that allows you to turn that steering wheel more easily."

**On using oxygen ...** "In normal sleep, sensitivity to CO<sub>2</sub> is reduced so that we all shallow breathe during REM sleep. Elevated CO<sub>2</sub> during REM sleep lets people stop breathing or slow breathe until the oxygen saturation levels get so low that the brain alarms to say, 'Wake up!' From that standpoint, supplemental oxygen tends to be a poison to people with this problem who are not also getting ventilation at the same time, because they lose the low O<sub>2</sub> wakeup alarm."

**Hypoventilation? Obstructive Sleep Apnea?  
Different Tests, Different Treatment**

**Judith R. Fischer, MSLS, Editor, Ventilator-Assisted Living**

***People with neuromuscular disorders may be misdiagnosed and mistreated when they encounter breathing and sleep problems. Many general practitioners, and even some pulmonologists, neurologists and sleep physicians, may not fully understand respiratory insufficiency and physiology in this group.***

**Hypoventilation.** Generally, in people with neuromuscular disorders who are having breathing problems, the main problem is hypoventilation (underventilation) – not breathing deeply and/or often enough. Muscle weakness, scoliosis and/or chest wall stiffness make it difficult or impossible to fully inflate the lungs.

Hypoventilation results in an imbalance in the carbon dioxide (CO<sub>2</sub>) and oxygen (O<sub>2</sub>) exchange in the blood – too much CO<sub>2</sub> is retained, too little O<sub>2</sub> is taken in. Because hypoventilation usually first occurs during sleep and because several of the signs and symptoms overlap, it can be misdiagnosed as obstructive sleep apnea (OSA).

Although any trained health care professional can perform simple pulmonary function tests (PFTs) of breathing ability during an office visit, the tests are most likely to be performed by a pulmonologist, neurologist, nurse or respiratory therapist. The challenge lies in understanding the results of these tests in the context of a person with neuromuscular disease.

Two important measurements of your ability to breathe deeply are the forced vital capacity (FVC) and maximum inspiratory pressure (MIP or PiMax). The SNIP (stiff nasal inspiratory pressure) test has been shown, in some studies, to be a more sensitive test of respiratory muscle weakness, but it is not widely used in the USA.

Forced vital capacity measures the volume of air you can breathe in and then blow out quickly and completely through a device called a spirometer.

It should be measured in both the upright and supine (lying face-up) positions, because you can't breathe as efficiently lying down.

Another simple test that measures the strength with which you can breathe in is the MIP. A mouthpiece is attached to a negative pressure gauge via a narrow tube. With a noseclip pinching off the nostrils, you exhale and then suck on the mouthpiece as hard as possible; the gauge registers the pressure.

A result of <50% predicted FVC or a MIP <60 cm H<sub>2</sub>O may signal that it's time to get some assistance with breathing.

However, the most important factor in diagnosing hypoventilation is an elevated level of CO<sub>2</sub>(above 45 mm Hg). This can be measured invasively with an arterial blood gas (ABG) analysis or noninvasively using exhaled end-tidal CO<sub>2</sub> monitoring or transcutaneous CO<sub>2</sub> monitoring.

The pattern seen on an overnight oximetry tracing may also be helpful for identifying early hypoventilation often seen first during the deepest rapid-eye movement (REM) sleep stage.

**Signs and symptoms of nocturnal hypoventilation may include one or more of the following:**

- fatigue or exhaustion after normal activity;
- excessive daytime sleepiness;
- shortness of breath, breathlessness with minimal activity;
- claustrophobia or feeling that air in room is somehow bad;
- difficulty in speaking for more than a short time;
- quiet speech with fewer words per breath;
- inability to lie flat while awake due to shortness of breath;
- inability to lie flat during sleep/need to sleep sitting up (orthopnea);
- trouble falling asleep and trouble staying asleep;
- anxiety about going to sleep.

**Other signs and symptoms, which may also be seen in OSA, include:**

- excessive daytime sleepiness and need to nap during the day;
- nightmares, night sweats, bedwetting, or need to urinate frequently;
- morning headaches;
- restless/fragmented sleep with frequent awakenings;
- shallow breathing or cessation of breathing for 10 seconds or more;
- awakening from sleep with choking sensation;
- worsening mental status, impaired memory, concentration, cognition.

***Do not ignore these signs and symptoms hoping they will go away. They are serious. You may need evaluation and treatment immediately!***

The treatment for hypoventilation is NOT oxygen but assisted ventilation, generally at night, with a bilevel ventilator. Bilevel units that offer the S/T mode (the unit operates in a spontaneous –S– mode, meaning the user can spontaneously initiate each ventilator breath, but switches to a timed –T– mode, referred to as the backup rate, when breaths are not initiated by the individual) are recommended for people with neuromuscular disorders.

Bilevel ventilators provide pressure support ventilation which is achieved by the difference in two set pressures: IPAP (inspiratory positive airway pressure) and EPAP (expiratory positive airway pressure). The IPAP and EPAP pressure settings can be adjusted separately.

People with neuromuscular disorders have more trouble breathing in. They generally need IPAP that is set at least 5-10 cm H<sub>2</sub>O higher than EPAP and EPAP that is set at the minimum level. Higher EPAP makes it too difficult for them to exhale. “In my home care company, we start out people new to bilevel with ‘training wheels’ – a minimum span of 5 cm H<sub>2</sub>O. After they become acclimated to the treatment, we increase the span if the individual is more comfortable and/or needs more volume,” says Diana Guth, RRT.

For reimbursement of a bilevel unit in the USA by Medicare, the requirements are a diagnosis of a progressive neuromuscular disorder, absence of chronic obstructive pulmonary disease (COPD) or if present it does not significantly contribute to the individual’s respiratory limitations, and one of the following test results:

- FVC <50% of predicted,
- MIP <60 cm H<sub>2</sub>O,
- PaCO<sub>2</sub> arterial blood gas >45 mm Hg,
- Nocturnal SpO<sub>2</sub> (oxygen saturation) <88% for five continuous minutes while asleep.

**Obstructive sleep apnea (OSA).** Apnea is the cessation of airflow for more than 10 seconds. OSA occurs when tissues in the throat collapse, intermittently blocking airflow during sleep. Snoring is often a major indicator of OSA, but not always.

A sleep study (polysomnogram test or PSGT) is primarily used to determine and design treatment for individuals with OSA. A sleep study is not

absolutely necessary for the diagnosis in people with neuromuscular disorders but it may be helpful when first introducing the bilevel treatment.

The main breathing problem is almost always hypoventilation, although people with neuromuscular disorders early on may also have undiagnosed OSA. Most sleep labs are not equipped to measure CO<sub>2</sub> levels, and therefore cannot diagnose hypoventilation.

The standard treatment for OSA is continuous positive airway pressure (CPAP) to help keep the airway open or a bilevel unit without a backup rate.

Thanks to Josh Benditt, MD, University of Washington, Seattle ([benditt@u.washington.edu](mailto:benditt@u.washington.edu)); Peter Gay, MD, Mayo Clinic; Diana Guth, RRT, Home Respiratory Care, Los Angeles ([Diana@hrcsleep.com](mailto:Diana@hrcsleep.com)); E.A. Oppenheimer, MD, (retired) Los Angeles; and Jesper Qvist, MD, Respiratory Centre East, Copenhagen, Denmark ([jq@dadlnet.dk](mailto:jq@dadlnet.dk)), for their assistance.

## **Breathing and Sleep Problems in Polio Survivors**

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

*Reviewed by Nicholas S. Hill, MD, Tufts-New England Medical Center, Boston, Massachusetts*

It is critically important that polio survivors, especially those diagnosed with post-polio syndrome, obtain proper testing, diagnosis, and management of breathing and sleep problems. The problems may result from weak breathing muscles in the chest and abdomen (diaphragm and intercostals).

Pulmonary function tests (mostly noninvasive) can measure the strength of respiratory muscles and usually include a test for forced vital capacity (FVC) – the maximum amount of air that can be exhaled. This test is typically administered when a person is sitting in the upright position, but *also* should be administered when a person is lying down (supine position). People who may not experience breathing problems sitting up may find themselves struggling to breathe when lying down. This problem is known as orthopnea, and a marked decrease in FVC while lying down indicates that the diaphragm is weak.

The late E.A. Oppenheimer, MD, a pulmonologist with years of experience treating polio survivors, described it, “As one ages with the late effects of polio, respiratory muscle strength may decrease. This may be particularly evident when you lie down, because in this position, the diaphragm has to work harder both to pull air in and also to push the intestines and other abdominal organs which are out of the way when one is upright due to gravity.”

Polio survivors also may have obstructive sleep apnea (OSA), central sleep apnea (CSA), a combination of OSA and CSA, and/or hypoventilation (an imbalance in the gas exchange in the lungs due to too little breathing - carbon dioxide builds up and oxygen drops). In obstructive sleep apnea, the upper airway collapses and blocks the flow of air so the person stops breathing periodically. These cessations of breathing are known as apneas and hypopneas. An overnight sleep study may be helpful to confirm the presence of OSA if it is suspected, based on snoring and daytime sleepiness.

Most sleep laboratories are set up to detect obstructive sleep apnea, for which the appropriate treatment is continuous positive airway pressure or CPAP, but not to measure carbon dioxide levels directly, which would be the best way to detect hypoventilation, for which the appropriate treatment is bilevel or positive pressure ventilation. The test for a high carbon dioxide level would be to measure arterial blood gases (requiring blood to be drawn via needle from an artery – usually in the wrist) or to monitor end-tidal or transcutaneous CO<sub>2</sub>, which is usually not done. However, sleep labs look for sustained drops in oxygen saturation during sleep that would be indicative of hypoventilation and then can proceed to additional testing to confirm hypoventilation, if needed.

When a bilevel device, a form of positive pressure ventilation, is prescribed, it will provide two levels of pressure; inspiratory (IPAP) to help blow air into the lungs, and expiratory (EPAP) to help keep the airways open and to blow off CO<sub>2</sub>. These pressures can be set and adjusted separately. IPAP settings are higher than EPAP settings, with a span of at least 8 to 10 advised for adequate breathing assistance, e.g. IPAP of 14, EPAP of 4. Bilevel units with a backup rate are recommended for people who may not be able to initiate a breath on their own, particularly at night. The bilevel device can provide timed backup breaths.

It is important for individuals to understand that polio survivors do not “forget to breathe” when they become overly fatigued. What happens is that the muscles become too weak to



move sufficient air, especially during sleep. Post-polio syndrome can also sometimes cause central sleep apnea which causes the brain to temporarily “forget” to signal breathing muscles to take a breath. This is evident during a sleep study when there is no chest wall movement for at least 10 seconds, indicating that the individual is not breathing and is apneic. Bilevel ventilation administered using a nasal or face mask can help with either problem.

Although a polio survivor may not have breathing or sleep problems when initially diagnosed with post-polio syndrome, periodic testing is important because such problems may develop over time or a change of breathing machine and/or settings may be warranted. (See *Tests for Breathing Problems If You Have a Neuromuscular Condition*.)

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

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