

Rancho Los Amigos Post-Polio Support Group
Newsletter Articles About Oxygen and Other Pulmonary Issues

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Hypoventilation? Obstructive Sleep Apnea? Different Tests, Different Treatment

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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 (CO₂) and oxygen (O₂) exchange in the blood; too much CO₂ is retained, too little O₂ 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 (sniff 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 mouth-piece as hard as possible; the gauge registers the pressure.

A result of <50% predicted FVC or a MIP <60 cm H₂O 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 CO₂ (above 45 mm Hg). This can be measured invasively with an arterial blood gas (ABG) analysis or non-invasively using exhaled end-tidal CO₂ monitoring or transcutaneous CO₂ 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 H₂O 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 H₂O. 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 H₂O,
- PaCO₂ arterial blood gas >45 mm Hg,
- Nocturnal SpO₂ (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 CO₂ 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.

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We have had many favorable comments on Dr. Oppenheimer's article on the dangers of oxygen usage that we printed in our August 2000 newsletter. Some individuals asked for additional information. They wanted to know more about the differences between COPD and post-polio. A few people seemed to be saying, "I feel better with oxygen. Why shouldn't I continue to use it?" Dr. Oppenheimer's follow up, and the other articles that appear in this newsletter may answer these questions.

A Follow Up on the Dangers of Oxygen

By E.A. Oppenheimer, MD

Physicians usually see two types of pulmonary impairments. The most common is Chronic Obstructive Pulmonary Disease (COPD), i.e. emphysema, asthma, chronic bronchitis, etc. COPD is a condition where the lungs or airways are damaged but the respiratory muscles function normally. The second is hypoventilation (underventilation) due to neuromuscular disease, i.e. polio, Muscular Dystrophy, ALS, etc. In neuromuscular hypoventilation the lungs are healthy, but weakened respiratory muscles impair the movement of air in and out of the lungs.

With COPD the problem is with oxygen exchange. The damaged lung tissues interfere with the transfer of inhaled oxygen into the bloodstream. The problem with post-polio and other neuromuscular diseases is the inability to move air (oxygen) effectively. Since COPD is so common, many physicians are used to treating a low oxygen saturation (even when there is an elevated CO₂) with oxygen, not considering the possibility that the problem is truly hypoventilation rather than COPD. Although modern blood gas equipment often automatically calculates the alveolar gas equation, many MDs and RTs no longer think in these terms and don't remember when or how to use this information.

Both COPD and hypoventilation due to neuromuscular disease result in a decrease of arterial oxygen levels as the problems get worse. If the doctor or respiratory therapist uses the "alveolar gas equation" to check the arterial blood gas (ABG) data in COPD patients it will show a wide alveolar to arterial oxygen difference due to the lung damage (ventilation:perfusion mismatch). But in post-polio hypoventilation the alveolar to arterial oxygen gradient is normal -- the calculation using the alveolar gas equation shows that all of the drop in oxygen is due to underventilation, due to the increased alveolar CO₂. This is often referred to as CO₂ retention.

Most physicians know that you need to be careful with oxygen administration in COPD. Too much can turn off respiratory drive and result in death. The body recognizes the higher oxygen level and tells the breathing muscles to slow down or breathe shallower. It's as if the nervous system is saying, "You've got plenty of oxygen. Slow down. You don't need that much." But COPD patients need some oxygen supplementation because of their problems with oxygen transfer.

Dr. Peter Gay (Pulmonary physician at the Mayo Clinic) published a review of similar problems when oxygen is given to people with neuromuscular disease. Already weakened respiratory muscles will be getting the wrong signals. The respiratory drive will turn off and death can result.

If COPD patients are given assisted ventilation without oxygen, the low oxygen problem persists. If a post-polio patient with low oxygen saturation is given mechanical ventilation, the oxygen level returns to normal without adding oxygen. This is because the post polio problem is underventilation. These patients respond to assisted ventilation alone. So why give the wrong treatment and risk a bad outcome? The answer is: You shouldn't! You should use mechanical ventilation to correct the underventilation, and avoid mixing up treatment of post-polio and COPD!

Post-Polio Breathing and Sleep Problems

By Judith R. Fischer and Joan L. Headley

New breathing and sleep problems in aging polio survivors can be insidious and often not recognized by either polio survivors themselves or their health care professionals. Polio survivors who were in an iron lung or barely escaped one during the acute phase should be aware of potential problems and educate themselves in order to avoid acute respiratory failure. Polio survivors who did not need ventilatory assistance during the acute phase may also be at risk for respiratory failure, and should likewise be aware of problems with breathing and sleep.

Simply stated, the major problem for polio survivors is hypoventilation (commonly referred to as underventilation). Not enough air reaches the lungs, producing an imbalance in the gas exchange in the blood: too little oxygen, too much carbon dioxide (CO₂) or hypercapnia. Hypoventilation is caused by one or a combination of the following: chest wall deformities such as scoliosis, respiratory muscle weakness due to the poliomyelitis, and sleep apnea (an interruption of breathing during sleep) which can be central, obstructive, or mixed and occurs in the general population as well.

Other factors contributing to a polio survivor's problems are a history of smoking, obesity, lung disease, and diminished vital capacity (VC). As aging occurs in anyone, vital capacity (VC) diminishes, but this decrease in VC is more serious in an aging polio survivor with limited musculature remaining to produce adequate ventilation.

Many polio survivors had impairment of the diaphragmatic and intercostal muscles, and the normal changes due to aging may cause them to lose VC at a greater rate, thus exacerbating the development of hypoventilation.

Signs and symptoms include: fatigue, daytime sleepiness, morning headaches, need to sleep sitting up, sleep disturbances (including dreams of being smothered, nightmares, restless sleep, interrupted sleep), snoring, poor concentration and impaired intellectual function, shortness of breath on exertion, 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, use of accessory muscles to breathe, and a weak cough with increased susceptibility to respiratory infections and pneumonias.

Polio survivors experiencing more than one of the above signs and symptoms should seek a respiratory evaluation by a pulmonologist, preferably one experienced in chronic neuromuscular disorders. (If your pulmonologist is unfamiliar with post-polio, you may wish to provide copies of pertinent literature, or offer to connect him or her with a physician in the I.V.U.N. Network.) Pulmonary function tests should include at least: VC, air flow, MIF (maximum inspiratory force) and MEF (maximum expiratory force), and ABG (arterial blood gases). A sleep study should also be considered. However, strong anecdotal evidence from polio survivors indicates that a sleep study may miss detection of hypoventilation. A sleep study is designed to detect sleep disturbances such as sleep apnea; it does not measure CO₂.

Management of breathing and sleep problems can be achieved largely through the use of nocturnal noninvasive ventilation in the form of mouth intermittent positive pressure ventilation, nasal intermittent positive pressure ventilation (a variety of commercial and custom nasal/face masks are available - see References) and negative pressure body ventilators. However, polio survivors may find themselves gradually extending periods of ventilator use. In some cases, as a last resort, invasive tracheostomy positive pressure may be necessary.

Medical literature regarding the management of breathing problems often includes a warning about the use of oxygen therapy. Respiratory failure in polio survivors is usually due to hypoventilation which can be aggravated by the short and long term use of oxygen. When the brain senses an excess concentration of CO₂ in the blood it instructs the body to increase breathing, ridding it of the CO₂ and increasing the oxygen. When oxygen is supplied, the mechanism in the brain detecting the CO₂ eventually turns off. Hypercapnia is best treated with assisted ventilation.

Polio survivors with different problems, such as COPD or pneumonia may benefit from short term oxygen therapy under careful supervision.

Thanks to Edward A. Oppenheimer, MD and John R. Bach, MD, for reviewing this article.

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Post-Polio Sleep and Breathing Problems: Assisted Breathing Aids

By Judith R. Fischer, Editor, IVUN News

People with neuromuscular disease such as post-polio often need help with breathing because the respiratory muscles are weakened or paralyzed. Often the chest wall is stiff and inelastic, and if scoliosis is present, the work of breathing may be further compromised. The inability to produce a good cough can lead to an increased incidence of respiratory tract infections and pneumonias. Sleep apnea may also occur. Underventilation is not uncommon and assisted breathing through mechanical ventilation can help people prevent respiratory failure. Oxygen is not the preferred treatment for underventilation and may exacerbate the condition. The following descriptive terms may be helpful.

CPAP - Continuous Positive Airway Pressure: Air flows continuously into the airways via the nose with use of a nasal mask. CPAP keeps the airways open, but does NOT assist inspiratory muscle activity directly NOR does it assist gas exchange in the lungs. John Bach, MD, describes it as “breathing with one’s head out of the window of a car going 60 mph.” Higher pressures make exhaling uncomfortable and difficult. CPAP is primarily used to treat obstructive sleep apnea, and thus is normally used only at night during sleep. CPAP units are not ventilators.

BiPAP - Bi-level Positive Airway Pressure: BiPAP refers to both a method of breathing and a machine. BiPAP units also continuously deliver air, but the inspiratory pressure can be adjusted separately from the expiratory pressure. BiPAP units are also used with a face or nasal mask or nasal pillows, and like CPAP, used mainly at night. (Like CPAP, BiPAP is not for 24-hour use.) However, BiPAP can only deliver a certain amount of pressure that may not be enough for people with respiratory insufficiency and underventilation. BiPAP is registered to Respironics, Inc. There are many other bi-level positive airway pressure support devices on the market, but the only one that can be truly called BiPAP is from Respironics.

Ventilators - Volume: Volume ventilators deliver a pre-set volume of air via nasal/face mask, nasal pillows, or tracheostomy tubes. These machines can deliver much more air than BiPAP units, and thus enable deeper breaths for improved coughing and air stacking. They may be necessary for people with poor lung elasticity and stiff chest walls, when BiPAP is not enough. Volume ventilators, though larger, heavier, and more expensive than bi-level units, are quieter, have more alarm features, and can be used for 24 hours. The most commonly used volume ventilators in the U.S. are the LP6 Plus, LP10, and new Achieva TM from Mallinckrodt, and the PLV-100 and PLV-102 from Respironics.

Ventilators - Next Generation: A new generation of ventilator technology has produced the LTVT™ series from Pulmonetic Systems, Inc., and the UniVent® Eagle® from IMPACT Instrumentation. These new ventilators are compressorless and run by turbines. They are very small - about the size of a laptop computer - and lightweight, about 13 lbs, but more expensive.

What is an interface? In reference to assisted breathing equipment, an interface is what connects the tubing from the ventilator or CPAP/bi-level unit to the person using it, either through nasal or facial masks, tracheostomy, lipseal, or mouthpiece.

Oxygen therapy - A useful analogy (from Lisa S. Krivickas, MD, Spaulding Rehabilitation Hospital) in regard to people with respiratory failure from neuromuscular disease is that the lungs are like a deflated balloon that is not strong enough to inflate. To inflate the balloon, mechanical assistance to force air into the balloon is needed. Blowing oxygen across the mouth of the balloon (the equivalent of using supplementary oxygen delivered by nasal cannula) will do nothing to inflate the balloon.

Assisted Coughing - Manually assisted coughing involves another person administering a thrust to the chest and abdomen of the individual with neuromuscular disease immediately after that individual takes a big breath. Mechanically assisted coughing can be performed with the In-Exsufflator Cough Machine (J H Emerson Co., Cambridge, Massachusetts). The In-Exsufflator applies positive pressure to provide a deep breath in, then shifts rapidly to negative pressure to create a high flow out, as with a normal cough. A newer model of the In-Exsufflator will be unveiled in 2000.

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Oppenheimer, E.A. (2000) Oxygen is NOT for hypoventilation in neuromuscular disease. IVUN News, 14 (1), 4. Copyright 2000 by Gazette International Networking Institute (GINI)

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Gonna Get Some of My Life Back... OH YEAH!

By Lili Guerry

My second sleep study last night using a BiPAP was Very Successful. During my first study I was monitored for eight hours, but only slept for the last three. There was enough data to show I needed help, but they wanted more time recorded while I was asleep. I took two Ambien with me to the center to take if needed.

Last night I had a wonderful male respiratory therapist. He was excellent with teaching me about equipment, discussing dream sleep and other stuff I did not know. He tried to get me to sleep in

total darkness, but I'm not ready for that yet, after polio and having many night lights and lamps on for years. Once when my husband went on a trip to Florida I had all my lights on, and a neighbor accused me of having an all night party. She wondered where all my guests parked.

After the therapist had me all wired for sound and everything was hooked up, he suggested I try to sleep first without taking Ambien. I went into a deep sleep within 30 minutes on my back. I haven't slept on my back in years - so long I don't remember. This started at approx 10 p.m. I awakened at 4 a.m., refreshed, wide awake and ready to eat. My voice was strong, and I had no headache, or muscle pain in either leg. My husband arrived at 5:45 a.m., as he too was anxious to hear what happened.

My friends, it is everything I prayed for and so much MORE!! The therapist found my window and then he got the BiPAP set with correct pressure for me. My breathing became as effortless as sliding down a velvet hill, naked. Just the BEST!!! What a gift!!! I shall be getting so much of my life back, with less fatigue, good deep REM healing sleep. The unit they prescribed for me can even be hooked up to a cigarette lighter plug. We can travel while I sleep in the back seat with BiPAP, and awaken refreshed upon arrival.

In my honest opinion, every polio survivor should have a sleep study done if you used any breathing equipment, or had any breathing difficulty, when you first had polio. It is vital for us to keep a good check on this area for help. Study muscle groups and how they affect breathing. With the net, Lincolnshire Library, and many, many polio sites, there is no excuse for ignorance. Education is KEY!

I have enclosed a site I have been reading from about the respiratory system. It's good information. Maybe others can send other sites as well. We must educate ourselves well about the late effects. I pester my doctors (in a friendly way) but I am very upfront with them and PPS. Gather material, print and keep it in all medical files for your doctors, dentist and your own self. Pass out this material freely. If you happen to run into a stiff shirt doctor don't give up. My pulmonologist was convinced I had cancer because of my left rib cage pain. It's all gone for now. I told him No! This is PPS. Listen to your body. It speaks to you and me and will not lie.

I am dancing in circles of joy now. I can "sing out" once more. Raise some hell and have some fun. I don't want to sleep my life away from so much fatigue. No wonder I felt depressed. But I had to experience it to know this is the correction for my problem. By the way, my respiratory therapist showed me that with the CPAP it's like exhaling through a cloth. PPSers need BiPAP, not CPAP. Fight for it. I tell you true and you can believe it. I would never lead you wrong.

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My Ventilator Story

By Richard Daggett

I contracted polio in 1953. Within twenty-four hours of entering the hospital I was given a tracheostomy and placed in a tank respirator. I could not move, swallow, or breath. Ten months after onset I was free from any ventilator, my tracheostomy was closed, and I was walking short distances. Over the next thirty or so years I functioned very well and maintained an active lifestyle.

In 1984 I experienced a sharp drop in my pulmonary capacity. My vital capacity went from about 1,100 ml to less than 800 ml, and has continued to decline. This is a result of what is commonly called “post-polio syndrome”. My CO₂ rose dramatically and my blood oxygen fell to a dangerous level. After trying several mouthpiece/nasal ventilator options with little success I made the decision to have another tracheostomy. It was my decision, and I have no regrets.

My upper extremity strength is limited, and I could never manage to put on or remove the mouthpiece/nasal apparatus by myself. There were too many straps and snaps. This has not been a problem with the trach positive ventilator. I can connect and disconnect the ventilator, and get in and out of bed by myself. I also feel very strongly that I am getting better ventilation.

There are some negatives. I have to clean the tracheostomy site daily, and I need suctioning sometimes. I average about once a day. Sometimes I will go two or three days between suctioning, and sometimes I may be suctioned three times in one day. If I have a severe respiratory infection I need to be suctioned more often, but I feel that congestion is a lot easier to manage with a trach. Suctioning feels weird, but it doesn't hurt. The trach has also changed the way I dress (I can no longer where a tie). But I remain active, and I have never felt shackled by my tracheostomy.

Having a tracheostomy is not an easy decision. It is not for everyone. For me, and for quite a number of others, it was the right decision. If you are experiencing sharply reduced pulmonary capacity, and are facing the possibility of a tracheostomy, don't feel like it's the end of world. Actually, for me it has been no big deal.

When your eyes don't see the fine print any more you wear glasses. If your hearing is diminished you would probably use hearing aids. If your legs become weaker, and walking is a problem, you start using a scooter or wheelchair. Making the decision to use a ventilator is much the same. You do it because it allows you the freedom to remain active. It's that simple.

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Oxygen is NOT for Hypoventilation in Neuromuscular Disease

By E. A. Oppenheimer, MD, FCCP

If progressive respiratory failure occurs in people with neuromuscular disease, an abnormal nocturnal oximetry study is often an early indication that hypoventilation is occurring. There are significant periods of decreased oxygen levels in the blood or hypoxemia during sleep when lying flat, in addition to decreases in vital capacity (VC), maximum inspiratory force (MIF), and maximum expiratory force (MEF). Decreased oxygen saturation (SaO₂) combined with increasing carbon dioxide (CO₂) retention or hypercapnia are the hallmarks of hypoventilation. This is sometimes called ventilatory pump failure, due to the weakened respiratory muscles.

Patients with neuromuscular diseases who are developing progressive respiratory failure due to respiratory muscle weakness will die unless mechanical ventilation is used. The rate of progression is often hard to predict. Some patients seem suddenly to experience life-threatening hypercapnic respiratory failure. They may not have been aware of gradually increasing symptoms and signs, particularly since they are often not physically active and are often not being regularly monitored with simple pulmonary function tests.

Administering oxygen does not provide assistance to the weakening respiratory muscles, but gives both the patient and the doctor the false impression that appropriate treatment is being provided. While in fact, hypoventilation is mistaken for an oxygen transfer problem. Indeed, administering oxygen can mask the problem. Also there is a danger of causing respiratory depression by giving oxygen. Oxygen is **not** the treatment for hypoventilation. It will improve the SaO₂, but not the hypoventilation and may increase the danger of dying of sudden respiratory failure.

In hypercapnic respiratory failure due to hypoventilation, the SaO₂ falls due to the rise of the CO₂. The alveoli in the lungs (tiny gas exchange units) should clear most of the CO₂ out with each breath. Instead, with hypoventilation, CO₂ accumulates and thus there is decreased room in the alveoli for oxygen. When mechanical ventilation using room air is provided, it lowers the CO₂ in the alveoli, corrects the SaO₂, and rests the respiratory muscles. The ventilator should be adjusted to achieve a normal SaO₂, on room air. If oxygen is being administered, one cannot use noninvasive oximetry to tell whether enough assisted ventilation is being provided; repeated arterial blood gas specimens (ABGs) would be needed.

When there is respiratory failure in neuromuscular patients (ALS, post-polio, SMA, muscular dystrophy, etc.) who have no additional pulmonary disease that impairs oxygen transfer, the ventilator set-up is adjusted to:

- be comfortable for the patient;
- achieve SaO₂ of 95% or higher on room air

- (this can be measured with a finger-sensor oximeter);
- assist the patient to effectively cough and clear secretions;
 - provide improved oral communication (if vocal communication is possible).

It has been common for people using noninvasive nasal ventilation (NPPV) with a bi-level positive pressure unit to use inadequate settings; frequently, they are not monitored with clinical evaluation and oximetry. The EPAP (expiratory) is often set too high -- usually it should not be higher than 3-4 cm H₂O; the IPAP (inspiratory) is set too low - usually it needs to be 12-16 cm H₂O and adjusted to achieve an oxygen saturation of 95% or higher.

Some situations may require administering oxygen, such as pneumonia due to infection or aspiration. If this occurs in patients with respiratory muscle weakness and hypoventilation, then it is important to provide both assisted ventilation and supplemental oxygen, and use ABGs to monitor them.

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Editor's Note: The *IVUN News* is an excellent resource for pulmonary information. We recommend that you subscribe to this publication if you have respiratory problems related to polio.

Personal Respiration Story

By Judith Hughes

In October 1998, I began thinking I needed a ventilator and saw a pulmonologist about it. During that initial visit I told him I'd been having a great deal of fatigue - mental and physical - and had to lie down at least 12 - 16 hours each day in order to function. I was unable to cook, dress myself, bathe regularly, go out, or visit with friends.

I told him I got particularly tired when I talked or sang, and that I'd frequently catch myself not bothering to breathe at all for long periods of times.

I also told him my spine no longer supported my upper body, that I had 100 degrees of lordo-scoliosis from polio at age 2. And I went on about spinal surgery in 1994 that had only been partially successful. Finally, I told him that an unintended secondary effect of this surgery was that my spinal curve had been shifted upward and I found myself with a concave chest. Preoperatively, it had been much more convex - like a woman's chest ought to be.

I remember the above because I wrote all this down and gave it to the pulmonologist. I also gave him information about polio ventilation. He sat and read it all, and then told me he wanted me to have a sleep study. He was treating quite a few other aging polios and talked to me about sleep apnea.

I had a sleep study. It showed I was apneaing all night long and my oxygen saturation level would go down as low as 25% during the night. He recommended I use a BiPAP (bi-level positive air pressure) machine at night. I got the BiPAP and used it religiously. Did I feel better? I thought I did. I was happy to have it. It seemed to relax me.

But I continued having apneas. I started fiddling with the inspiration pressure - turning it up. I told this to my doctor and he said that I was using the machine like a ventilator. He encouraged me to up the inspiration pressure.

My apneas continued throughout the next year. I used the machine faithfully, but kept getting weaker and less mentally alert. I spent at least two full days a week quietly in bed.

In November 1999, I saw my pulmonologist and, again, told him I needed a ventilator. The BiPAP wasn't working. He arranged another BiPAP study. Maybe there was a problem with my pressure settings.

The second technician was a very competent fellow, and accurately titrated my BiPAP. He happily told me as he left that I wouldn't be having more apnea episodes. He was right, they did stop. But, to get this accurate titration, he reduced my inspiration pressure and raised my expiration pressure. I no longer had the "extra" air to help me breathe in, and I had to breathe out against a wall of air.

Then I really went downhill.

I got very weak and shaky. My face, hands, legs and feet were constantly bloated. I avoided people as much as possible. It was the winter I thought. I'm really weak in the winter. I stopped going out. I stopped getting dressed. I was good for about 4 hours a day, but then even 4 hours was too many. I skipped many baths, skipped just about everything. I avoided people because people tired me, and I thought I'd die if I got more tired.

The more tired I got, the more I wondered about the BiPAP. I was never out of breath, but my chest burned if I talked too much. My heart was racing with very little activity; there was a low-level of "agitation" throughout my whole body. I kept thinking, maybe a ventilator would help, but would I then go back to apnea? I didn't want to apnea, I didn't want to die.

You don't think very well when you're profoundly exhausted.

I started reading everything I could about ventilation and polio. I read that most people get ventilated during an emergency. They are intubated because pulmonologists in emergency services just don't know much about non-invasive ventilation. Intubation is quick, it works, and it saves lives. I determined I wasn't going to let that happen to me.

I decided I was going to get a ventilator. My whole plan involved letting my pulmonologist see me tired. The two afternoons before my appointment with him, I "visited" with friends, talking for about 4 hours each afternoon. Then to really exhaust myself, I took a bath before seeing the pulmonologist.

When I saw him I was in a sorry state. I hadn't let myself get that tired in a long time - I knew better. My pulse was 138. That scared me. It scared the doctor, too. A quick ABG was done. I could hardly talk. He said to me, "What is it you want? Do you want me to do a trach?" "No," I replied, "I want to use a ventilator non-invasively." A hour after I got home I got a call about when could my ventilators be brought over.

So, I got a vent. Big, ugly PLV-100. My brain was so tired I was appalled by the number of dials and readouts on the thing. I've come to appreciate the features. The feature I liked the best was the screaming the machine does if I block off the passage of air - I realized this machine was going to easily take care of my apneas.

But after about 4 days on the vent, I felt even more tired. The timing and pressures just weren't right for me.

One exhausted night, while watching the Horse Whisperer - a very long, long film - I set about adjusting a couple of dials. My body seemed to want a deeper breath, and fewer breaths per minute than the technician thought might be right for me. While Robert Redford calmed the traumatized horse, I got my inspirations and expirations just right.

Bottom line: once the ventilator pressure/timing settings were right for me, I began to come back to life. Water began pouring out of me on the fourth and fifth day of use: my face and body were

no longer bloated, nor were my legs. My heart calmed down and my body feels quiet again. I sleep quite well, and haven't been aware of any apneas.

One of the first things I did when I came back to life was to get my hair cut, buy some new make up, and now I know I need some new clothes.

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