

## Airway Clearance and Lung Volume Recruitment for Individuals with Neuromuscular Disease

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To maintain airway clearance, individuals with neuromuscular disease (NMD) must have an effective cough, which requires a high volume and flow of air, but often these individuals have an ineffective and weak cough. This can be due to several factors, including small lung volumes, weak inspiratory and expiratory muscles, stiff inelastic chest wall, and difficulty with coordination or weakness of the glottis, i.e., the voice box. The glottis must close tightly as the pressure builds up in the chest before a cough.

During an upper respiratory tract infection, when a strong cough is needed most, lung volumes are lower and respiratory muscles are weaker. Since ineffective coughing will result in retained lung secretions, a common cold could lead to severe respiratory illness, such as pneumonia, requiring an emergency department visit, an ICU admission, or even an endotracheal

tube or tracheostomy. Obviously, for a quality of life and the opportunity to remain at home, strategies to improve cough and airway clearance are critical.

Noninvasive airway clearance methods can be highly effective in improving cough capacity and lung volumes. One of the easiest methods is lung volume recruitment (LVR), commonly referred

to as breath-stacking. The most common approach uses a simple hand-held resuscitation bag modified with some added tubing and one-way valves (Figure 1), but LVR can be done with a mouthpiece, if one has a chair-mounted, volume-targeted ventilator, or with glossopharyngeal (frog) breathing.

To perform LVR effectively, the muscles of the lips, mouth and voice box need to be reasonably strong to seal completely around the mouthpiece and to stack breaths. This is called "Active" LVR. If the muscles are weak, alternatives to provide volume include using a well-sealed full face mask or a non-vented nasal mask. The seal is created by the mask and the valves in the tubing, not by the lips and voice box.

The mouthpiece is placed just inside the sealed lips and held in place while the bag is squeezed and air fills the lungs. After one squeeze, the volume is held by the voice box and another squeeze of volume is added to (stacked on top of) the first. In this way the

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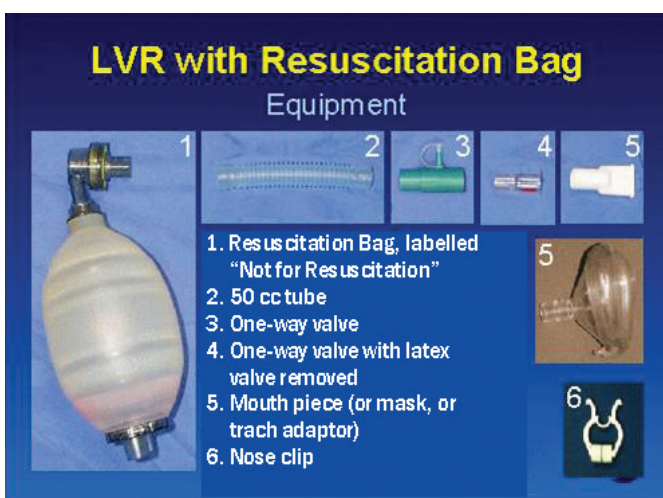


Figure 1. The usual equipment includes a one-way valve with the valve removed (clear valve labelled 4.) and the inner screen remaining to prevent the valve from entering the airway should it become dislodged. A full face mask can be used if more severe weakness is present.

## From Around the Network

Judith R. Fischer, MSLS, IVUN Information Specialist, [info@ventusers.org](mailto:info@ventusers.org)

**International Ventilator Users Network's mission is to enhance the lives and independence of home mechanical ventilator users and polio survivors through education, advocacy, research and networking.**

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### Home Mechanical Ventilation Guidelines and Consensus Statements

**“Domiciliary Non-Invasive Ventilation in Adult Patients”** is the title of a consensus statement from the Respiratory Network of the Agency for Clinical Innovation in New South Wales, Australia. The comprehensive 200-page report was produced by a working group led by Amanda Piper, PhD, Senior Clinician Physiotherapist, Royal Prince Alfred Hospital in Sydney, Australia. Chapters include general and disease-specific recommendations for mechanical ventilation, slowly progressive as well as rapidly progressive neuromuscular disorders, hypercapnic vs. non-hypercapnic central sleep apnea, transition from nocturnal to continuous NIV use and from paediatric to adult care, and palliative and end-of-life care. [www.health.nsw.gov.au/gmct/respiratory](http://www.health.nsw.gov.au/gmct/respiratory)

**“Guidelines for Non-Invasive and Invasive Mechanical Ventilation for Treatment of Chronic Respiratory Failure”** was produced by a group from Germany led by Prof. Dr. med. Wolfram Windisch. The guidelines detail the technical setup of mechanical ventilation, diagnostic tests, transition from hospital to home, use of NIV in both obstructive and restrictive diseases, and end-of-life care. *Pneumologie* 2010;64:640-652. [www.thieme.de/pneumologie](http://www.thieme.de/pneumologie)

**“Home Ventilation” guidelines** from the Canadian Thoracic Society (CTS) will soon be available online. Co-chairs of the CTS Home Mechanical Ventilation Committee are Douglas McKim, MD, The Ottawa Hospital Rehabilitation Centre, Ontario, and Jeremy Road, MD, Gordon and Leslie Diamond Health Care Centre, Vancouver, British Columbia. Check the CTS website: [www.respiratoryguidelines.ca](http://www.respiratoryguidelines.ca). ▲



*Joan Gillespie, polio survivor, uses both mouthpiece and mask ventilation during the day and a customized full face mask at night. She had polio as an infant and used an iron lung until she was 8 years old. Joan was able to breathe independently until her middle years, but in 1979 she began using the iron lung to sleep in at night. Joan successfully converted to NIV in the late 1980s. Joan lives at home with her husband and is active in the community, volunteering at the local hospital.*

## Victorian Respiratory Support Service

Anne Duncan, RN, VRSS Outreach Coordinator, Heidelberg, Victoria, Australia, [anne.duncan@austin.org.au](mailto:anne.duncan@austin.org.au)

The Victorian Respiratory Support Service (VRSS) is funded by the state of Victoria to provide respiratory assessment and review, and ventilators, related equipment and care to people in Victoria who need a ventilator to support their breathing. Referrals are made to the service by general practitioners and specialist physicians.

Currently, the VRSS has approximately 640 patients using ventilators. Most patients use noninvasive ventilation (NIV), via a mask or mouthpiece, but a few people need ventilation via a tracheostomy tube or electrical pacing of the diaphragm. We use predominantly bilevel pressure ventilators. Volume ventilators and equipment with back-up battery support are used as required.

Our team chooses the appropriate equipment and adjusts ventilator settings on at least two occasions initially and is available to provide ongoing support and adjustment of equipment at subsequent follow-up. Masks used by our patients include nasal and full face masks from a variety of manufacturers and a customised nasal mask (made by our occupational therapist for use only with volume ventilators). Unfortunately, VRSS does not have enough funds to loan out secretion clearance devices, except for suctioning units to those with tracheostomies.

Most patients who use NIV are managed by VRSS, but a few are managed using a share care model. In this model, VRSS supplies the ventilator and associated equipment, provides annual home visits for equipment maintenance, mask fitting and other equipment issues. Ongoing respiratory care is provided by the referring (public) hospital. Each group exchanges the outcome of visits in order for all

parties to have up-to-date records of the patient's care and status. Paediatric patients are managed by the Royal Children's Hospital (RCH) until the age of 18, at which time there is a staged transition from the RCH to the VRSS.

Sleep studies require a written referral from a respiratory physician. The VRSS conducts diagnostic studies before initiating NIV, but sometimes commences NIV symptomatically, and then conducts a sleep study to ensure adequate ventilation, as part of the follow-up. Treatment review studies will be done if the patient has any symptoms of inadequate ventilation, such as fragmented sleep, morning headaches or daytime sleepiness.

Most of our patients live in their own homes, but about 4 percent live in nursing homes and other community residential facilities. Diagnoses include obesity hypoventilation syndrome, motor neurone disease, spinal cord injury, muscular dystrophy and other neuromuscular diseases, post-polio syndrome, central hypoventilation syndrome, kyphoscoliosis, combined COPD/hypoventilation and bronchiectasis. We also have a small number of patients who use NIV as a bridge to lung transplant. ▲

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lungs and rib cage are expanded toward more normal volumes that could not be achieved otherwise. The bag can be squeezed by a caregiver or if capable, by the individual (Figure 2). The LVR technique is recommended to be performed at least once in the morning and once in the evening but preferably more often. Ten to 15 full lung inflations, with periods of rest to prevent hyperinflation, should



Figure 2. Individuals with spinal cord injury (left) and ALS (right) performing LVR on themselves. The full face mask was necessary due to weakness of oral and throat muscles.

*“Think of lung volume recruitment as range of motion therapy for the respiratory system, just as a physiotherapist would perform on an arm or leg which had become stiff.”*

be achieved each session but it is not known what the optimum frequency should be.

The individual usually begins from a full breath, but starting from an empty or a more relaxed volume is sometimes easier when first learning. It may be possible to take only one breath at a time or several depending on how stiff and how small the lungs and rib cage are.

The natural lung volume that is exhaled from full to empty is called vital capacity (VC). The largest volume that can be held with LVR is the maximal insufflation capacity (MIC). The best measure of effective LVR is

the MIC-VC difference (the largest volume held with LVR minus the vital capacity). This difference is the volume that drives a stronger cough and is, in fact, the most critical value in determining the ability to maintain noninvasive ventilation, even for 24 hours a day.

To increase the expiratory force of a cough or the cough peak flow (CPF), a manual abdominal thrust (manually-assisted cough) and, when necessary, mechanical generation of positive and negative airway pressures (mechanical insufflation-exsufflation, such as the CoughAssist®) can also be used. Cough peak flow can be measured in litres per minute (L/min) using a peak flow meter. To prevent respiratory complications, an effective CPF should be at least 270 L/min.

Many patients have achieved both an increase in cough capacity and an improvement in their VC, even in progressive NMD. One patient with Duchenne muscular dystrophy showed better sleep quality, weight gain, less shortness of breath, and improved blood gases, VC, MIC and CPF as a result of the regular performance of LVR. A tracheostomy and percutaneous endoscopic gastrostomy (PEG) tube that had been recommended elsewhere were still not required six years later.

Any improvements in the flexibility of the lung and rib cage may mean that weak muscles do not have to work quite so hard to inflate the lungs. In addition, ventilators may be equally effective at lower pressures or more effective at current pressures. This could improve mask fit and comfort while ensuring adequate ventilation. (If patients are not regularly performing LVR, they should

Complete instructions from Dr. McKim's team on the use of LVR, frog breathing and assisted cough techniques, "Respiratory Care Protocols: Spinal Cord Injuries and Diseases; Unit 3 - Respiratory Interventions: Lung Volume Recruitment and Assisted Cough" are available online: [www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=1](http://www.irrd.ca/education/slide.asp?RefName=e2r4&slideid=1)

▷ ask their pulmonologist whether it would be helpful.) It is important to measure VC, MIC and CPF both naturally and with LVR.

Although serious consequences of LVR are very rare, a few patients have experienced collapsed lungs as a result of this technique, but in 17 years of caring for hundreds of patients, our team has not seen a single individual with a serious consequence of LVR. Initial stretching of the respiratory system could produce some discomfort, similar to range of motion therapy. Individuals with spinal cord injury who may be prone to low blood pressure could feel very light-headed and should perform LVR in a more reclined position, such as in bed, as a part of their morning and evening routines.

Lung volume recruitment is an incredibly safe, inexpensive and effective way to increase cough capacity, lung capacity and maintain respiratory health. It is a critical therapy for individuals with respiratory muscle weakness and limited cough capacity who are at risk for acute respiratory failure. ▲

#### Reference

McKim, D. Keeping ventilated and 'at-risk' patients out of the intensive care unit. *Can Resp J*, 2008;15 (Supplement C); 9-10. [www.pulsus.com/journals/JnlSupToc.jsp?sCurrPg=journal&jnlKy=4&supKy=445](http://www.pulsus.com/journals/JnlSupToc.jsp?sCurrPg=journal&jnlKy=4&supKy=445)

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To register for this webinar, visit [www.passy-muir.com/eventwebinar](http://www.passy-muir.com/eventwebinar). ▲



Jack Rushton

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See "Learning to Use a Speaking Valve:"

[www.ventusers.org/edu/valnews/val\\_24-4aug10p1-6-7.pdf](http://www.ventusers.org/edu/valnews/val_24-4aug10p1-6-7.pdf)

# Ask The Experts

**QUESTION:** *Physicians are required to indicate a “diagnosis” for their patients from pre-defined categories for insurance purposes. My new pulmonologist, who struggled to find a plausible one for my constellation of respiratory problems (polio, iron lung at first, then night-time ventilation since 1952), finally chose chronic respiratory failure and added a note about polio. Is there a more appropriate choice? Chronic respiratory failure sounds dreadful.*

**ANSWER:** Brenda J. Butka, MD, Respiratory Care & Pulmonology, Vanderbilt Stallworth Rehabilitation Hospital, Nashville, Tennessee

“Chronic respiratory failure” does sound dreadful, but so does “congestive heart failure” – and there are treatments for both, and neither means the heart or lungs are just about ready to stop!

“Respiratory failure” is a perfectly accurate description that applies to anyone who uses oxygen or has elevated carbon dioxide or is very short of breath from any chronic respiratory condition. Other diagnoses that I might use, along with this one – not instead of it – might be “restrictive lung disease,” or “disorder of the diaphragm,” or “obstructive sleep apnea,” or “central apnea,” and, of course, I would reference polio.

Your doctor sounds like he or she is accurately assessing the facts of your physiology, which is the first step to appropriate treatment. Just remind yourself, as I occasionally have to remind my patients, that you are not any different after this box was checked than you were before. This is not news – just a label. But a label that is helpful to your doctor as he works with you on managing your condition.

**ANSWER:** Oscar A. Schwartz, MD, FCCP, FAASM, Sleep Disorder Specialist and Medical Director of the Barnes-Jewish West County Hospital Sleep Disorder/EEG Center, St. Louis, Missouri

This concern has troubled polio survivors and physicians for years. There are multiple terms used to describe the respiratory problems faced by individuals requiring respiratory support devices. The term “respiratory failure” has always been the least complimentary. No one appreciates being referred to as a “failure.”

The late effects of polio (ICD-9M 138) and hypoventilation (ICD-9M 768.09) in medical terms also describe chronic conditions but appear to be less negative. Coding is dependent on regional practices and also on the acceptable terminology necessary to qualify for the equipment required and not the disease state in particular.

Unless the physician has the acceptable code, the ventilator or other needs may be rejected by the insurance carrier. I agree the coding and how the health care profession makes reference to conditions is confusing and, at times, appears to be insensitive. Remember, it is only a code! The real mission is to use the respiratory support device to allow for the most productive lifestyle possible. ▲

## REMINDER! Get Your Annual Seasonal Flu Shot

Seasonal flu can be a very serious illness. Over a 30-year period from 1976 to 2006, estimates of annual flu-related deaths in the United States range from a low of 3,000 to a high of 49,000. The composition of this year’s shot is the same as last year, but it should be taken again. Get your flu shot as soon as the vaccine is available. See [www.flu.gov](http://www.flu.gov).

**OCTOBER 22-27, 2011. Neuromuscular Respiratory Medicine: Theory, Practice and Hands-on Demonstrations.** CHEST, American College of Chest Physicians Annual Conference, Honolulu, Hawaii. [www.chestnet.org](http://www.chestnet.org)

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In the afternoon, the course provides an opportunity for pulmonologists and respiratory nurses and therapists to see and touch portable ventilators, masks and other interfaces, and assisted cough devices.

For information on course fees: <http://2011.accpmeeting.org/program/additional-saturday-courses>

**MARCH 15-17, 2012. 13th International Conference on Home Mechanical Ventilation,** Barcelona, Spain. Sponsored by Journées Internationales de Ventilation à Domicile (JIVD) and European Respiratory Care Association (ERCA).

*Abstracts due January 15, 2012.* Contact Brigitte Hautier, JIVD, c/o BAL Congrès, 7 rue Belfort, 69004 Lyon, France, +33 (0)4 78 39 08 43; [brigitte.hautier@free.fr](mailto:brigitte.hautier@free.fr); [www.jivd-france.com](http://www.jivd-france.com).

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