

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.

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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

- ▷ 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

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