Addendum to Breathing Problems of Polio Survivors

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EQUIPMENT

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Polio survivors and people with other neuromuscular diseases may need help with breathing because respiratory muscles are paralyzed or weakened. The chest walls of polio survivors may be stiff and inelastic, and, if they have scoliosis (curvature of the spine), breathing may be more difficult.

Problems with breathing usually are noticed first during sleep and can be due to underventilation, to sleep apnea (obstructive, central, or mixed), or to a combination of both. To determine the best equipment for treatment, it is important to determine the correct diagnosis, either through pulmonary function tests, pulse oximetry during the night, and/or a sleep study.

The following descriptions may be helpful when considering assisted breathing options.

Interface

An interface is what connects the circuit or tubing from the equipment to the person using it. This connection can be invasive through a tracheostomy, or noninvasive via nasal or facial mask, a lipseal, or a mouthpiece.

CPAP – Continuous Positive Airway Pressure

When using a CPAP machine, air flows continuously into the airways via the nose with the use of a nasal mask to keep the airways open. CPAP 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." CPAP is primarily used to treat obstructive sleep apnea and is normally used only at night during sleep. CPAP units are not ventilators and will not be useful if the sleep or breathing problem is due to underventilation.

Newer units, known as auto-titrating positive airway pressure units, automatically adjust the pressure and provide varying levels of pressure throughout the night based on the individual's needs. They may also be used diagnostically. The pressure is generally lower than that of standard CPAP units. The AutoSet® T from ResMed (www.resmed.com) is an example.

BiPAP – Bi-level Positive Airway Pressure

BiPAP machines continuously deliver air, as do CPAP machines, but the inspiratory pressure can be adjusted separately from the expiratory pressure. These respiratory assist devices are usually prescribed for people with mild to moderate inspiratory muscle weakness and underventilation. It is recommended that the initial inspiratory pressure be set at 8-10 and gradually increased if necessary. The expiratory pressure on set-up should not be higher than 3-4, but may be adjusted lower. Like CPAP, BiPAP units are used with a face or nasal mask or nasal pillows, mainly at night. BiPAP units also compensate for mask leaks better than volume ventilators *(see following).*

There are many other bi-level positive airway pressure ventilators on the market, but the only one that can be truly called BiPAP® is registered to Respironics, Inc. (www.respironics.com).

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 breathing, coughing, and air stacking. Volume ventilators may be the most beneficial equipment for people with more severe respiratory muscle weakness, poor lung elasticity, and stiff chest walls. Volume ventilators, though larger, heavier, and more expensive than bi-level ventilators, are quieter and have more alarm features. The most commonly used volume ventilators in the USA are the LP6 Plus, LP10, and Achieva® series from Puritan Bennett (www.mallinckrodt.com), and the PLV®-100 and PLV®-102 from Respironics.

Ventilators – Next Generation

A new generation of ventilator technology has produced the LTV™ series from Pulmonetic Systems, Inc. (www.pulmonetic.com). This new ventilator is "compressorless" and is run by turbines. It is very small – about the size of a laptop computer – and lightweight, about 13 pounds, but more expensive than volume ventilators. Some models provide both pressure and volume modes, while the latest and simplest model (LTV800™) provides only volume ventilation.

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Assisted Coughing

Most often the lungs of polio survivors are healthy. However, the inability to produce a good cough can lead to an increased incidence of respiratory tract infections and pneumonias. Thus, clearing secretions is extremely important. Manually assisted coughing involves another person administering a thrust to the chest and abdomen of the individual immediately after that individual takes a big breath. Mechanically assisted coughing can be performed with the CoughAssist™ from J.H. Emerson Co. (www.coughassist.com) that 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.

References

Bach, J.R. (1999). *Guide to the evaluation and management of neuromuscular disease.* Philadelphia, PA: Hanley & Belfus, Inc.

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OXYGEN USE

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Are there circumstances when oxygen use might be appropriate for a person with respiratory muscle weakness due to neuromuscular or musculoskeletal disease?

There are at least six situations in which using supplemental oxygen is warranted.

For people who are not using any type of ventilator:

1. Oxygen might be used if the individual does not want to use any type of assisted ventilation but wishes to have symptomatic relief without improving survival; this is often called palliative care.

2. Supplemental oxygen is justified if the arterial carbon dioxide (CO_2) is repeatedly not elevated, indicating that there is no hypoventilation, but the oxygen saturation (SaO_2) is 88% or lower and the arterial oxygen (PaO_2) is 55 mm Hg or less. Adjusting the added oxygen to achieve oxygen saturation of 90-95% should be on the advice of one's physician. He or she may prescribe it only at night or whenever the oxygen saturation is 88% or lower. Sometimes the cause for this is difficult to identify but can occur more often at higher altitudes, such as Denver or Mexico City, than at sea level. At higher altitudes, atmospheric pressure is reduced and this decreases the availability of oxygen.

For people who are using a ventilator (noninvasively or via tracheostomy) to successfully treat hypoventilation resulting in normal arterial CO_2:

3. Oxygen might be needed if there is also a chronic lung or heart problem such as COPD, pulmonary fibrosis, or heart failure.

4. Oxygen might be needed if there is an acute lung problem, such as pneumonia, until this resolves.

5. Oxygen might be needed if one is living (or visiting) at higher altitudes. If the ventilator is adjusted properly so the arterial CO_2 is normal, but the high altitude causes the oxygen saturation to drop to 88% or lower and the arterial PaO_2 is 55 mm Hg or less, then supplemental oxygen is justified – again, adjusting the added oxygen to achieve an oxygen saturation of 90-95% following the advice of one's physician.

A variation of this, often at high altitudes, might be that the night-time adjustment of the ventilator, despite all attempts, is not optimal (perhaps due to leaks), so the physician may suggest adding supplemental oxygen rather than considering tracheostomy.

6. In air travel, the airplane often flies at about 30,000 feet, with the cabin pressure adjusted to between 5,000 to 8,000 feet. This is equivalent to being at high altitude. The arterial oxygen would drop by at least 16 mm Hg, compared to the sea level value. A person who usually only uses night-time ventilation may need supplemental oxygen during air travel. A physician would need to advise, arrange this, and determine the oxygen flow (usually available either with 2 or 4 liters per minute). Some people need to be sure a small portable oxygen tank will be available in order to leave one's seat and use the toilet.

The general guideline is true – *oxygen is not the right treatment for hypoventilation due to respiratory muscle weakness.* In fact, using oxygen rather than assisted ventilation can result in serious complications. However, in the situations described above, using oxygen can be quite reasonable and important. Each individual needs to discuss this with a physician to get the best advice. ■