BiPAP/Ventilation

Assistance for Breathing Problems
Pulmonary health is a significant issue. It is important for ALS patients to learn about mechanical ventilation by getting good information and talking to peers. Then, when the situation presents itself, a person can make an informed decision. This report presents the identifying signs of pulmonary insufficiency and a variety of available options.

The Respiratory System
The primary functions of the respiratory system are to bring oxygen into the lungs, transfer the oxygen to the blood, to expel the waste product called carbon dioxide, and to help regulate acid-base balance. Oxygenated blood travels from the lungs through the pulmonary veins and into the left side of the heart, which pumps the blood to the rest of the body. Oxygen-depleted, carbon dioxide-rich blood returns to the right side of the heart and is pumped through the pulmonary artery to the lungs, where it picks up oxygen and releases carbon dioxide.

Because the lungs have no muscles of their own, the work of breathing is done primarily by the diaphragm and, to a lesser extent, by the intercostal muscles (between the ribs). During forced or labored breathing, other muscles in the neck, chest wall, and abdomen also participate. As the diaphragm contracts, it moves down, enlarging the chest cavity. This reduces pressure in the chest and air rushes into the lungs to equalize the pressure. The diaphragm then relaxes and moves up; the chest cavity contracts and raises the air pressure. Air is pushed out of the lungs because of their elasticity. The intercostal muscles participate in this process, especially if breathing is deep or rapid. When everything is working well a person hardly notices that he is breathing.

Symptoms
Breathing problems can sneak up on ALS patients because they may not be easily identified. Is your answer “Yes” to any of these questions?

- Are you experiencing increased fatigue?
- Do you frequently awaken with a headache?
- Are you having problems sleeping?
- Do you need to use additional pillows when sleeping?
- Do you sleep better in a recliner or chair than in your bed?
- Do you have a poor cough or difficulty clearing secretions?

If your answer was “Yes” to any of these questions, then it is probably time for a breathing evaluation. If your answer to all of the questions was “No”, it is still important to educate yourself about possible breathing difficulties in ALS patients so that you will be able to recognize a problem if it does occur.
Self-Help

It is critical to have an effective cough. The strength of a person’s cough can be assessed with a Peak Flow Meter (cost $15-$20). The individual takes a deep breath and then coughs forcefully into the Peak Flow Meter. A “peak cough flow” reading of less than 180 liters/min. indicates a weak cough, according to John R. Bach, MD. A cough can be improved by:

- Using huff coughing (not the high force cough, but repeated gentler huffs to bring up secretions)
- Using the breath stacking technique (while using a mechanical breathing aid, hold your breath after one or more inspiration cycles and add the air from the next inspiration cycle), or with frog breathing (GPB). Examples of mechanical devices used for this are an Ambu-type resuscitator bag and a volume ventilator.
- Manual cough assistance methods (having someone push on your upper abdomen as you cough)
- Devices to improve cough and clearance of secretions, such as Emerson’s CoughAssist device, formerly known as the In-Exsufflator [J.H. Emerson Co., Cambridge, MA (800) 252-1414]

If breathing muscles are weak because of ALS do not use up your energy on exercising to improve their strength. It is unlikely to help. However, the ability of the lungs to expand should be maintained. Using a chest inflation machine, learning how to take a deeper breath, and glossopharyngeal breathing (GPB) – sometimes called “frog breathing”, can help maintain the ability of the lungs to expand. GPB can augment the volume of air you breathe in; it is an important survival skill to learn if your respiratory muscles are weak.

Use of a Continuous Positive Airway Pressure (CPAP) machine may also require increased work for the expiratory breathing muscles. The CPAP was developed during the 1980s to keep airways open for people with sleep apnea. During the 1990s cyclical inspiratory pressure was added to this equipment and the Bilevel Positive Airway Pressure (BiPAP) came into use. The BiPAP assists inspiration. Bilevel equipment is made by a number of manufacturers. When respiratory muscles are weak due to PPS, a bilevel device would usually be better and safer than CPAP even if a sleep study shows sleep apneas.

Avoid poor nutrition. Unless your muscles get the required amount of nutrition as well as oxygen, they will function less effectively and any weakness will become increasingly noticeable. If a person has lost weight and become frail, finding ways to build up his nutritional status will help, in terms of his immune status and muscle function, as long as he doesn’t become overweight.

Sleep Study

Sleep aggravates weak breathing muscles and developing respiratory failure. Thus more breathing problems occur at night. If daytime breathing tests are within normal range, a simple over-night oximetry study can be done at home (this continuously records pulse and oxygen saturation). Sometimes a more expensive and elaborate over-night sleep study is needed. During this procedure a patient is connected to many measuring devices and when indicated, assistive breathing equipment may also be used to decide its effectiveness and to determine the proper settings. During REM sleep (deep “rapid eye movement” sleep) accessory breathing muscles and other voluntary muscles become so relaxed they are sometimes referred to as paralyzed. That is why breathing problems are first noticed during REM sleep because at that time breathing becomes completely dependent upon the patient’s diaphragm, which may be weak due to ALS.
Bulbar Impairment
When an ALS patient has bulbar impairment there is a triple threat: weak respiratory muscles, a poor cough (which can lead to development of infection and pneumonia), and the risk of food aspiration.

Choices
Choosing Home Mechanical Ventilation (HMV) is conditional upon:

- Quality of life is worthwhile.
- Physician presents the options.
- Resources are available (particularly care giving).
- Social support is available for HMV (hand in hand with medical support).
- Benefits outweigh the burdens.

Most people in the United States think of a ventilator as evil equipment. So without any advance education most would choose to withhold using a ventilator when deciding in advance. However, 95% of those who are already on HMV, want to remain on it. So it is important for everyone to learn about HMV by getting good information and talking to peers. Then, if the situation presents itself later, a person can make an informed decision. An ALS patient who is having breathing problems has four choices:

1. He/She can decline assisted ventilation and use only palliative care.
2. He/She can use only non-invasive Nasal Positive Pressure Ventilation (NPPV), such as BiPAP.
3. He/She can use assisted ventilation only for acute episodes such as pneumonia.
4. He/She can use whatever ventilation support is needed for long-term survival, including trach positive pressure ventilation if needed.

By providing experience with noninvasive ventilatory equipment (such as BiPAP) before a crisis occurs, the patient will then have the ability to survive an acute respiratory infection if it does develop. And, this also provides a “hands on” trial of the equipment to help decision making (like a “test drive”). The goals of HMV are to integrate the individual back into the community, avoid creating a hospital ICU environment, and avoid technological entrapment.

When to Start Nasal Positive Pressure Ventilation (NPPV)
If a person needs to use more pillows to elevate his head in bed, or prefers to sleep in a chair or a recliner, or his legs begin to swell, these symptoms may be enough motivation to begin using ventilatory equipment. Physiologic tests should confirm that chronic respiratory failure is developing. Any one of the following confirmatory tests justify starting NPPV:

- A person’s vital capacity (VC) is less than 50%, although Dr. Oppenheimer said that he might start NPPV if the VC is 60%; or
- The maximal inspiratory and expiratory pressure readings are less than 50 cm. of water; or
- If nighttime oximetry shows evidence of under-ventilation, with periods of low oxygen saturation of 88% or less for at least five minutes.

The old criteria used abnormal Arterial Blood Gas (ABG) values as a determinant. Regulatory agencies prefer tests like ABG tests which cannot easily be gamed (data manipulated to justify
NPPV equipment). But ABG values are often abnormal “too late”, just when a respiratory failure crisis is occurring.

Oxygen
Initially the goals of NPPV are to relieve symptoms, normalize sleep, provide experience with NPPV as hours of use gradually increase (see if you like it), prepare for the ability to survive acute respiratory infection, and maintain blood oxygen (O2) saturation levels at 95% without added oxygen. When the ventilation device is working properly the carbon dioxide (CO2) is eliminated and the patient’s oxygen level is high; with improper ventilation the patient’s oxygen level drops. If a person is not getting enough breathing support the CO2 level will climb and leave less room in the lungs for oxygen, so the O2 level falls. If a person wants to monitor oxygen levels himself, Dolly Bodine, RRT, a representative of Mallinckrodt (Puritan-Bennett) and Dr. Oppenheimer recommend the Nonin brand’s ONYX finger pulse oximeter (it is small, accurate, and less expensive): 800-356-8874. Often this is not covered by health plans or Medicare.

What blood oxygen levels are normal? Readings of 95% (at sea level but lower at higher elevations) and nocturnal desaturation episodes as low as 90% may be normal. Nocturnal oxygen saturation of 88% or less is abnormal. When blood oxygen saturation levels are below 85% you enter the danger area; below 75% you are in the disaster area.

“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.”

“Administering oxygen does not provide assistance to the weakening respiratory muscles. It 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 (oxygen saturation level in the blood), but not the hypoventilation and may increase the danger of dying of sudden respiratory failure.”

After an abnormal sleep study shows decreased oxygen levels, some physicians naturally want to prescribe oxygen. A study by Peter Gay, MD (Mayo Clinic) concluded that the use of oxygen could turn off a person’s respiratory drive (and death can occur as a result).

Care Plan for using Nasal Positive Pressure Ventilation
Considerable expertise is required to set up NPPV equipment effectively and comfortably. Frequent follow-up is needed, similar to that of a person who has a trach. The interface (mask, nasal pillows, etc.) should be checked and the effectiveness of assistive ventilation determined. Objective assessment includes examining the patient, checking the equipment, making sure that the oximetry is 95% or better (without added oxygen), and sometimes doing overnight oximetry monitoring. Regular follow-up is important.

With a bilevel ventilator, initially the inspiratory positive air pressure (IPAP) setting on the machine may be at 8 to 10 cm. of water, but this setting often needs to be gradually increased to between 14-16 cm. Persons with scoliosis may require an even higher setting. Initial expiratory positive air pressure (EPAP) setting will be about 3-4 cm. and usually will not be changed.

Safety issues are critical. The person using ventilatory equipment must learn all the equipment problems that could possibly develop and know how to fix them. Regular safety drills should be done with all caregivers so they will have the same knowledge. If these safety issues are neglected, a person using
ventilatory equipment leaves himself open to possible catastrophe. An Ambu-type resuscitator bag should be available.

**Tracheostomy or Non-Invasive Ventilation**

A tracheostomy should be considered if NPPV fails, if the person has a problem with secretions and/or prefers a trach, or when long-term survival is important. Safety may be better with a tracheostomy when daily 18 to 24-hour ventilator life support is needed. Does a trach ventilator provide the most effective ventilation? The BiPAP also provides effective ventilation, but neither is perfect. Dr. Oppenheimer advises that if a trach works for you keep it, no matter what any physician tells you. Doctors' opinions will vary on this matter. If you have a trach you should also have a good suction machine. More suctioning may be required during the first year while the trach heals and matures. (Medicare may cover the monthly rental charge of about $30.00 until the machine is paid off in a year or two.)

**In the Future**

There are some people who function better with volume ventilators and nasal masks rather than bilevel equipment. New equipment will be combining the advantages of both types of equipment. Smart BiPAP machines are now being developed. These machines will have the ability to recognize what a person needs and automatically adjust the settings accordingly. New alarm features on the BiPAP will compare favorably to the safety features of a volume ventilator. Improved nasal and full-face interfaces (masks) are becoming available. A straight or angled, or lip seal mouthpiece is useful for many, and is important to have in case of nasal congestion during a common cold or allergy.

**Conclusion**

There are many benefits from Nasal Positive Pressure Ventilation:

- It provides muscle rest and recovery at night.
- It helps reset the CO2 sensitivity in the respiratory control system.
- It probably improves respiratory mechanics.
- It helps with periods of low oxygen and helps prevent acidosis.
- It improves the quality of sleep.
- It can improve cough and swallowing.
- It improves long-term survival.

Fortunately the outlook today for mechanical ventilation is better than it was ten years ago because now there are more physicians and respiratory therapists who are familiar with the new noninvasive ventilatory equipment that is available.

This article was reprinted with permission from Edward Anthony Oppenheimer, MD, FCCP. It is based on a report written by Mary Clarke Atwood with editorial assistance from R. Daggert, V. Duboucheron, and E. A. Oppenheimer, MD, FCCP. Minor modifications were made to the article so that it applies particularly to PALS, since it was originally developed for polio survivors. The original report was based upon a presentation by Dr. Edward A. Oppenheimer, MD, FCCP, to the Rancho Los Amigos Post-Polio Support Group on March 24, 2001. He had just returned from the Eighth International Conference on Home Mechanical Ventilation held in Lyon, France. Dr. Oppenheimer is Associate Clinical Professor of Medicine, University of California, Los Angeles (UCLA). He retired after 31 years with Kaiser Permanente Medical Group where he organized and coordinated the Kaiser home ventilator program. Although Dr. Oppenheimer is no longer seeing people in office practice and does not have an office referral, he will try to respond to comments and questions emailed to him at Enopp@UCLA.edu.