Non-Invasive Positive Pressure Ventilation AKS BiPAP in ALS
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ALS can affect breathing, coughing and swallowing by weakening the muscles that serve these functions. When this happens assisted ventilation can be an important way to improve the quality and length of life.

The type of ventilation used most often for this purpose is known as BiPAP, which stands for Bilevel Positive Airway Pressure. Other names for this device include non-invasive positive pressure ventilation (NPPV) or bi-level ventilation and respiratory assist device (RAD) Therapy is usually started when the symptoms of decreased respirations are first noticed. These may include a sense of shortness of breath, which is typically the worst when exercising or lying flat on the back. Difficulty removing the by-product of metabolism, carbon dioxide, may also occur, usually during the deepest of sleep states. Symptoms of carbon dioxide retention include a sense of chronic fatigue, excessive daytime sleepiness or headaches upon awakening.

The Pulmonary and Circulatory Systems

To understand the use of bi-level ventilation, a basic understanding of the pulmonary and circulatory systems is helpful. The lung is an organ that has many large airway passages that transfer air to small air sacs, alveoli, deep within the lungs. The right side of the heart pumps blood through the lungs, that ends up in tiny capillaries where oxygen can move from the air cells into the blood. The oxygen is carried by red blood cells that return to the left side of the heart and are pumped throughout the body. Red blood cells release oxygen when they reach different organs and pick up carbon dioxide, which returns through the veins back to the heart and then to the lungs where it is exhaled.

There is a feedback loop involving the brain that controls the rate of respiration. When the body is producing carbon dioxide in normal amounts, for example at rest, the breathing rate is slow. Conversely, when the body is using oxygen more quickly, such as while exercising, it also produces more carbon dioxide and this causes breathing to increase in depth and rate. This change in breathing keeps the oxygen and carbon dioxide at consistent levels.

Muscles of Respiration

The primary muscle of respiration is the diaphragm, a large muscle situated under the lungs. When the diaphragm contracts during inspiration, it moves downward and opens up the lungs, causing air to flow into the lungs. When the diaphragm relaxes, it moves upwards and pushes air, now containing carbon dioxide, back out of the lungs. If the diaphragm becomes weakened in ALS, it impairs the ability to take a deep breath.

Other muscles involved in breathing are termed the “accessory muscles of respiration” and include the neck, chest, abdominal and back muscles. These muscles are used in breathing under
times of need such as intense exercise. When the diaphragm is weak, these muscles end up doing some of the work of breathing to help keep oxygen and carbon dioxide levels normal.

Use of accessory muscles may first become noticeable after eating or when lying down. During deep sleep, the accessory muscles “turn off” and the only muscle working is the diaphragm. Frequent awakenings could signal low oxygen levels if the diaphragm cannot carry the work of respiration by itself.

**Definition of NPPV or Bi-level Ventilation**

The term “BiPAP”, is derived from “bi” meaning two (as in inhalation and exhalation) and PAP meaning positive airway pressure (positive pressure means air is pushed into the lungs). We prefer the term non-invasive positive pressure ventilation (NPPV) for this type of device since BiPAP is actually only the name of the original bi-level machine. There are other machines now available that do the same thing.

NPPV designates two levels of positive pressure in the airways of the lungs. The machine applies a maximal amount of pressure directly into the airways when it senses that the patient is beginning inspiration (IPAP – inspiratory positive airway pressure). A second, smaller amount of pressure is exerted on expiration (EPAP- expiratory positive airway pressure). This lower amount holds the airways open and keeps the level of oxygen elevated to normal. When NPPV is set up on ALS patients, a “backup” breathing rate is usually set as well. If the breathing rate falls below a set level the machine can automatically give additional breathes.

**When Should NPPV Be Considered?**

Symptoms that may indicate the need for assisted breathing include:

1. Difficulty lying flat.
2. Awakening at night due to shortness of breath
3. Frequent nighttime awakenings. Patients suspect reasons other than breathing such as an urge to urinate or simple discomfort. However, if the diaphragm is not strong, abnormal oxygen and carbon dioxide levels can result in this symptom.
4. Excessive daytime fatigue and sleepiness due to poor nighttime sleep.
5. Not feeling refreshed and rested upon awakening.
6. Morning headaches. This occurs when carbon dioxide is retained in the blood.
7. Wild nightmares.
8. Night sweats.
9. Mental confusion.

**Testing to determine need for NPPV**

There are several tests that can monitor the function of the respiratory muscles.
1. Forced Vital Capacity (FVC) – This measures the amount of air that is exhaled after taking as large a breath as possible. The result depends on your size, since different people have different lung capacities. The test is usually performed sitting up. If the diaphragm is weak, the FVC decreases since the lungs cannot expand maximally. A value of less than 50% of the predicted volume for size and age usually correlates with the need for NPPV.

2. Maximum Inspiratory Pressure (MIP) – This measures the amount of negative pressure the diaphragm can exert against a pressure meter. It compares to the strength that the lungs can suck in. The MIP decreases when the diaphragm is weak and a value of less than –60 cm H2O (centimeter of water pressure) corresponds to the level where NPPV is considered.

3. Nighttime oxygen study (Nocturnal Oximetry) – This measures the amount of oxygen being carried in the blood. It is done by placing a device on the finger that measures oxygen all night while sleeping. This may be done in a specialized sleep lab or in the home (although many insurance companies will not pay for a home study). Oxygen levels of less than 88% of normal, lasting for 5 or more consecutive minutes are an indication for NPPV.

4. Polysomnography – This study is typically done in a specialized sleep lab where it can be observed by a sleep technician. It measures several functions including how air flows through the nose and mouth, oxygen saturation, heart and brain waves, and movements of the chest and body.

5. Arterial blood gas – This test involves placing a small needle into an artery near the wrist. It measures the exact amount of oxygen and carbon dioxide in arterial blood that has left the lungs. Carbon dioxide levels of more than 45 mm HG suggest that the lungs are not working hard enough to get rid of this gas, and indicate the need for NPPV.

**Benefits of NPPV**

NPPV is usually used at night for periods of 4 hours or longer. It aims to improve breathing at night, levels of oxygen and carbon dioxide, and quality of sleep. By improving sleep conditions, patients feel less daytime fatigue and sleepiness and improved mood. Morning headaches also resolve. If symptoms do not improve initially, they often respond to continued use of the equipment.

While NPPV is typically used during the night, it may be used during naps as well. Some patients use NPPV before meals to help improve appetite, or during periods when breathing is difficult. Patients may use NPPV for up to 24 hours during periods of respiratory illness such as pneumonia or permanently with severe muscle weakness.

**NPPV in Various Settings**

The NPPV should never be considered a deterrent to travel. With appropriate adaptors, inverters and batteries the NPPV may be used in a car or plane, on the back of a wheelchair, or in foreign countries.
Use of NPPV

A respiratory therapist with proper training and equipment can determine the correct pressures needed to deliver adequate amounts of air during each breath. A physician will order the NPPV and interface from a home medical equipment company, who will usually set it up in the home.

Initial Set-up

Expect the home care company to set up the equipment within a week after the order is received. Request that the set up be done in the home. Place the equipment in the area where the patient sleeps. Try the NPPV both while seated and lying down and make adjustments at the time of the visit. Set up in the home will usually take about an hour. There should be at least one other person attending the set up meeting. Be sure all questions are answered, instruction booklets are received and phone numbers obtained.

There are several different brands of NPPV, specifically BiPAP ST and Synchrony by Respironics, VPAP III STA by Resmed and Knightstar by Puritan Bennett. All orders should include a specific inspiratory and expiratory pressure and the backup respiratory rate. Asking the respiratory therapist about orders and settings is very appropriate. Oxygen may also be ordered with the NPPV, although most patients with ALS don’t need oxygen.

Make sure to clarify the following before the respiratory therapist leaves:

1. How to use the NPPV machine, and how to respond to alarms and error codes. An instruction booklet should also be provided.
2. What changes to breathing therapy the patient can and cannot make.
3. Putting the mask or similar device on and taking it off.
4. Care needed for the equipment including cleaning, filters, and special instructions.
5. Alternate power source instructions.
6. How to communicate if one needs help while on the NPPV. An alarm or a communication device may be considered

All patients respond differently to NPPV. Some will sleep the whole night right away while others will need to increase therapy gradually an hour each night or week. The goal is usually four or more hours per night. Whatever happens, therapy should never be painful or make shortness of breath occur. If this happens, call the respiratory therapist or doctor.

When the NPPV is received, it should be attempted every night. However, this is not always an easy task and getting to sleep may be difficult. Following are some suggestions to try:

1. Try using the equipment during the day, napping or watching TV.
2. Gradually increase the amount of time used at night.
3. Ask for advice from your home care respiratory therapist.
4. Talk to your doctor.
5. Ask your doctor about using sleep or relaxant medications.
6. KEEP TRYING; do not just stop using NPPV without informing the physician. Discuss problems with the home care therapist and or your doctor. You are not the first person to have problems with NPPV.

There are some occasions when NPPV is not tolerated or is inadequate, in these cases other equipment may be ordered including a volume ventilator, negative pressure ventilator, or pneumobelt.

**Interface to NPPV**

The facial interface you, your physician and the home respiratory therapist choose is of primary importance. There are basically four types of interfaces to NPPV, nasal masks and pillows, oral, and full-face masks. In helping choose the interface best suiting you, consider the following and discuss with the respiratory therapist and physician.

1. Do you breathe through your mouth or nose?
2. When you sleep does your mouth come wide open?
3. Are you claustrophobic?
4. Are you allergic to certain types of plastics?
5. Do you have sensitive skin?
6. Do you sleep on your side or back?
7. Do you read right before going to sleep?
8. Do you watch TV right before sleep or when you wake up in the middle of the night?
9. Do you move around a lot while sleeping?
10. Are you able to speak loudly enough to call for help if needed?
11. Do you have a lot of oral secretions?
12. Do you have nasal problems? i.e. deviated septum, postnasal drip, and allergies?

**Online purchases** of interfaces are possible. If you have Medicare insurance, interfaces will need to be billed by your home care company. With private insurance you may be able to purchase online and submit it to your carrier. Online purchases are usually less expensive, need a physicians order but care should be taken with regard to sizing and type. If at all possible, get the mask sized before ordering.

Note: Red, sore dry eyes may occur due to NPPV interface leaking air into the eyes. This is unacceptable and preventable with the correct type and size of interface. Notify the respiratory therapist and the MD as needed.

**Common Problems associated with NPPV**

**Skin Problems**
When initially using NPPV, understand that there should not be any skin problems and if there are, contact your doctor and/or respiratory therapist to get treatment and suggestions. There are some general rules that will help prevent skin problems:

1. A slight leak around the bottom of the mask is OK, but not into the eyes
2. Don’t over tighten the mask, keep as loose as possible
3. The marks on your face from the mask should go away after a couple of hours, if not the mask is too tight.
4. If you are trying to decide between two different sizes of the same mask, usually the smaller of the two is better.
5. Switch between two different masks frequently if a second mask is available.

Upper airway problems

NPPV compresses the room air and delivers flow and volume into the lungs. Frequently this additional air may cause upper airway problems including dryness of the nasal passages and throat and increased viscosity of mucous in the lungs. Other upper airway problems may include nasal stuffiness, runny nose, bloody nose, laryngospasm, and difficulty managing oral secretions.

Airway Dryness

Airway dryness is uncomfortable and may deter patients from using NPPV on a regular basis. Drinking plenty of fluids during the day and keeping water at the bedside may help. A heated humidifier lets air carry more moisture, but some patients do well with a cool humidifier. If a heated humidifier is already in use, it may be necessary to increase the temperature to increase the moisture level.

Note that adding humidity can cause water to “rain out” in the tubing. Water rain out occurs due to the cooling of the air from the humidifier. There are some humidifiers that adjust to the room temperature, decreasing the rain out. Other suggestions that may be tried to prevent rain out include wrapping the tubing, tucking the tubing under the blankets to keep it warm and a high pressure water trap. Be sure to empty the water from the humidifier, turn it off and disconnect the hoses during the day.

There are several over-the-counter creams that may be tried including Ayr, Nose Better, acid mantle cream and Roezit. Occasionally nasal saline sprays also work well.

The interface may need to be changed. For instance the nasal pillow type interface may dry out the nasal passages more than a nasal mask. A chinstrap may be needed to prevent air leaking back out of the mouth and a possible change to a full-face mask may be indicated.

Oral secretions

Patients who have weakness of swallowing may find the use of NPPV challenging. Medications that decrease saliva or thin mucous may help. Nebulizer treatments containing mucous thinners, high frequency chest wall oscillation (vest) therapy and a coffalator may prove valuable in removing mucous prior to going to bed. Sleeping with the head of the bed elevated helps keep secretions from pooling in the back of the throat. Other strategies include using stacked breathing with assisted cough and keeping a suction machine at the bedside.
Nasal plugging

Some patients may experience nasal stuffiness. This may be treated with nasal steroids, though these take several days to start working. Discuss this with the physician. Try to avoid over-the-counter sinus medications such as Afrin, which may cause rebound stuffiness. Some patients actually respond to a nasal strip like sports players use. These are inexpensive and lack side effects.

Runny Nose

Runny nose is caused by increased flow through the nose or lack of humidification. Time may allow the nose to adjust or a humidifier may help. Persons with allergies may find prescription allergy medications helpful.

Bloody Nose

A small amount of blood now and then is expected, usually from some drying of the mucous membranes covering the nasal passage. The use of blood thinners or aspirin-like medications may make this problem worse and it will be necessary to stop therapy until a physician has been notified.

Laryngospasm

In some patients, the larynx and vocal cords have a tendency to go into “spasm” as a result of being stimulated by the breathing device. This causes the sensation of shortness of breath or an inability to inhale or exhale. While this sensation may be frightening, laryngospasm resolves in a matter of seconds. Laryngospasm occurs in some ALS patients even without NPPV where it responds to breathing through the nose, swallowing or moving the head. NPPV sometimes can be a treatment for laryngospasm. When NPPV causes laryngospasm it is important to work with the physician and respiratory therapist. Medications, adjustments of flow rate and pressures may help.

Other Problems Associated with NPPV

Two gastrointestinal problems that may be caused or worsened by NPPV are acid reflux and stomach distention. Both of these problems need the attention of a physician.

Stomach Distension and Gas

Stomach distension due to air swallowing may occur from NPPV, especially if high pressures are needed. The air in the stomach may cause burping, bloating, and abdominal pain. Gas in the stomach can also cause shortness of breath if the stomach pushes on the diaphragm, making it harder for the diaphragm to move downwards during inspiration. This can be treated by slowing the flow rate or decreasing pressures on NPPV. If the patient has a stomach tube in place, opening the cap helps alleviate this problem. Preventing constipation may prove helpful, as will other distension management methods.
Acid Reflux or Gastroesophogeal reflux disease (GERD)

Acid reflux is a common problem with ALS patients. This may occur from the effects of changing pressures when there is a weak diaphragm. The diaphragm serves as one of the key control mechanisms for preventing stomach acid from flowing upward into the esophagus. Symptoms of GERD include heartburn, shortness of breath, a bad taste in the mouth, and nausea. Management may include antacids, acid reducers, and medications that speed up the emptying of the stomach.

Other strategies for management include:
1. Elevating the head of the bed during sleep
2. Avoid eating 3 hours before bedtime
3. Prevent overeating including large tube feedings
4. If on continuous tube feedings around the clock, discuss with the MD
5. Avoid irritating foods including caffeine containing foods, peppermint, high fat foods and alcohol.
6. Avoid tight nightwear.
7. Stop smoking.

Successful NPPV Follow-up

Once NPPV is being used successfully, it is still important to monitor progress. A physician might order nighttime oximetry testing from time-to-time to insure that oxygenation is adequate. Ventilator pressures and rates can be changed and rarely the additional use of oxygen may be added.

The respiratory therapist or physician can help if problems arise as a result of therapy. The home care company should routinely check the equipment for function, and the condition of tubing, filters and interfaces. If you are unsatisfied with home care, it is very important to communicate this with the company or the physician. Unfortunately, some patients end up not using NPPV therapy because of initial discomfort. Communication is essential to assure maximal benefit. There are many home medical equipment companies and most often another can be used if problems cannot be resolved.

Care and Cleaning of NPPV Equipment

All instructions for care and cleaning will be given verbally and the patient should receive a booklet on NPPV equipment including ventilator, interface and humidifier

NPPV
Clean with damp towel routinely. Change filters as needed, generally once per month. Do not place the ventilator on the floor to operate, as the air will then be pulled from the floor resulting in dirty filters more frequently. If airborne allergies are a problem, pollen filters may be available. Ask the respiratory therapist.
Humidifier
This should be cleaned with soap and water. Antibacterial soap is not necessary and lemon scented soap should be avoided. Some humidifies may be cleaned in the dishwasher, check the owner’s manual.

Always clean the humidifier daily putting fresh distilled water in each evening. Humidifiers are a great source for bacterial growth; fastidious cleaning is essential to prevent respiratory infection.

Interface
NPPV interfaces should generally be cleaned with soap and water once per week and wiped out daily. Tubing should be cleaned weekly. During a respiratory infection, clean interface daily. Headgear should be cleaned as needed by hand washing and hanging dry.