

Maintaining Good Breathing with ALS

ALS can weaken the muscles that control breathing. When this happens, patients may have a hard time moving enough air into and out of their lungs. This can result in breathing that is too shallow, especially at night. The lungs of ALS patients remain normal. It is the muscles that move the lungs that cause problems. Unlike patients with respiratory disease, the use of an oxygen tank is not recommended, and can sometimes make the problem worse.

If breathing is too shallow at night, you will not stay in a deep, restful level of sleep (REM sleep). You might be in a lighter level of sleep or you might even wake up. As a result of shallow breathing, you may experience some or all of the following problems:

Signs of Respiratory Problems

- Waking up in the morning feeling tired
- Difficulty staying awake during the day
- Waking up at night feeling like you were short of breath
- Waking up in the morning with a headache
- Feeling short of breath while lying flat in bed
- Difficulty with concentration or memory
- Shortness of breath on exertion
- Lack of energy or increased fatigue

How can your nighttime breathing be improved?

When needed, most ALS patients use a small machine to assist their breathing at night. This is called non-invasive ventilation (NIV) or non-invasive positive pressure ventilation (NPPV). One brand name is BiPAP[®] and it stands for Bi-level Positive Airway Pressure ventilation. NPPV will help you breath more deeply, and have a more restful, useful sleep. **It has been well documented that NPPV increases the length of life and the quality of life in people with ALS.**

How do I know if I need assisted ventilation?

There are a variety of tests that a Respiratory Therapist can use to determine how well you are breathing. These include using breathing tests to tell the strength of the diaphragm (the main muscle that controls breathing) or testing how much oxygen is present in your blood.

What is NPPV?

NPPV stands for “Non-invasive Positive Pressure Ventilation.” “Positive Pressure” means that extra air is passed by your nose or mouth so that when you take a breath, it becomes a bigger breath and you breath deeper.

There are different types of masks that can be used with NPPV. Some just fit the nose, while others fit the nose and mouth, or the entire face. You and your respiratory therapist can discuss which type might work best for you. Finding a mask with the right fit is essential. NPPV is usually used at night. It needs to be used for at least 4 hours during sleep to really help the night time breathing problems. Some people may choose to also use it during the day to make them more comfortable or to rest.

What are possible problems with NPPV?

Some patients experience facial discomfort from wearing the mask. Patients may choose to rotate the use of different masks, to avoid irritation in one area of the skin. Patients who have difficulty swallowing may have a hard time with NPPV because of a build up of mucous or saliva at night. Sleeping with the head elevated, and using certain mucus thinning medications, medications to dry saliva, or mechanical coughalators (a machine to help you cough up and remove mucus) prior to bed can help. Airway dryness can also be a problem with NPPV, if a humidifier is not added to the machine.

In Summary:

1. Tests done by a respiratory therapist and careful questioning of the patient can tell if it is time for non-invasive ventilation (NPPV) to help breathing at night.
2. Potential problems of NPPV can be handled with patience and practice.
- 3. Studies have shown that NPPV can increase a patient's length of life and give a better quality of life (especially increased energy) if used for at least 4 hours during sleep every night.**

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